



# Cardiology in the Young

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### O1-1

#### Quality of Life, Psychosocial Adjustment, Psychiatric Morbidity, School Performance, Physical Limitations and Social Support in Adolescents and Young Adults with Congenital Heart Disease: How these variables play together?

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**Objectives:** The aims of this investigation were to study Quality of Life (QoL), Psychiatric Morbidity (PM), Psychosocial Adjustment (PSA), School Performance (SP), Physical Limitations (PL), and Social Support (SS) of adolescents and young adults with Congenital Heart Disease (CHD).

**Methods:** 74 CHD patients, 41 male and 33 female, aged from 12 and 26 years (mean = 18,76 ± 3,86), The original cardiac malformation was cyanotic in 45 and non-cyanotic in 29. Participants were interviewed once on topics as social support, family educational style, self-image, physical limitations and emotional adjustment, were administered a standardized psychiatric interview (SADS-L) and filled self-report questionnaires on QoL (WHOQOL-BREF) and psychosocial adjustment (YSR and ASR). One of their relatives filled the observational versions of the same questionnaires (CBCL, ABCL). Full clinical and demographic history was collected.

**Results:** There was 23% lifetime prevalence of psychopathology and 51.4% of retentions in school (M = 1.74 year +0.86). There were no differences in QoL for severity or type of CHD, nor psychiatric diagnosis. Comparing our patients to healthy population, we found better social relationships (SR) (t = 2,333; p = 0,022) and environment (t = 3,754; p = 0,000) QoL. Patients' without pharmacological therapy revealed better QoL in SR domain (t = -2,226; p = 0,029). Being submitted to surgical procedures decreases physical (t = -1,989; p = 0,050), SR (t = -2,012; p = 0,048) and general (u = 563,000; p = 0,037) QoL and leads to more withdrawn PSA (u = 238,500; p = 0,012).

SS is very important in improving patients' physical (t = 3,287; p = 0,002), psychological (t = 3,094; p = 0,003), SR (t = 3,669; p = 0,000), environment (t = 2,725; p = 0,008) and general (u = 323,000; p = 0,005) QoL, but those with poorer SS had more withdrawn (u = 767,500; p = 0,005) and delinquent behavior (u = 745,000; p = 0,011). Patients' with PL showed worse physical (t = -2,910; p = 0,005) psychological (t = -2,046; p = 0,044) and general (u = 947,500; p = 0,001) QoL and more withdrawn PSA (u = 449,500; p = 0,016). Patients and relatives don't agree about gender expressions of PSA. Female patients refer more somatic complaints (u = 886,000; p = 0,021), anxiety/depression (u = 952,500; p = 0,003), aggressive behaviour (u = 999,000; p = 0,005), thought problems (u = 929,500; p = 0,005), but relatives think that boys, instead of girls, show more withdrawn (u = 341,500; p = 0,019) and aggressive behavior (u = 665,500; p = 0,050).

**Conclusions:** While CHD patients seem to be more prone to PM, bad PSA and bad SP, SS plays a crucial role in all variables and in resilience.

### O1-2

#### Sense of coherence in adolescents with congenital heart disease in the transition to adulthood

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**Introduction:** Some studies have found that the quality of life (QOL) of patients with congenital heart disease (CHD) is better than that of healthy peers. At first sight, this finding is counterintuitive. However, it is hypothesised that this better QOL in patients with CHD is explained by a higher sense of coherence (SOC). SOC is a key concept of the salutogenic theory, which explains how individuals manage stress and stay healthy. Indeed, previous research in various patient populations concluded that SOC is positively associated with QOL. In the

present study, we aimed (i) to test the hypothesis that the level of SOC in adolescents born with CHD is higher than in healthy peers, and (ii) to explore the association between SOC and QOL in patients with CHD.

**Methods:** This cross-sectional study was part of a 4-wave longitudinal study on transfer and transition in adolescents with CHD. A total of 429 adolescents (14–18 years) with CHD participated (response rate: 86%), 403 of which were matched on gender and age to healthy controls (1:1 matching). The level of SOC was determined using the 13-item version of Antonovsky's SOC-scale (SOC-13). All items are scored on a 7-point Likert scale (i.e., total scale score ranging from 13 to 91). Higher scores indicate a higher level of SOC. Overall quality of life was measured using a Linear Analogue Scale (LAS) ranging from 0 (worst imaginable quality of life) to 100 (best imaginable quality of life).

**Results:** The median score on the SOC-13 scale was 61 (Q1 = 53; Q3 = 71) in adolescents with CHD. This score was significantly higher than the median score of 52 (Q1 = 46; Q3 = 60) in healthy controls ( $Z = -9.503$ ;  $p < 0.001$ ). A Pearson's correlation showed that SOC was positively associated with QOL ( $r = 0.474$ ;  $p < 0.001$ ).

**Conclusions:** This study showed that, as expected, adolescents with CHD have a higher level of SOC. Furthermore, SOC was confirmed to be associated with QOL. Hence, the present study corroborates the hypothesis that patients with CHD can have a better QOL due to a higher SOC.

#### O1-3

##### **Transition from pediatric cardiology to GUCH – information from the Swedish National Registry SWEDCON**

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**Introduction:** SWEDCON, a national registry for congenital heart disease, has been created in Sweden and includes data from pediatric cardiology, GUCH and congenital cardiac surgery. The registry has only been in use for 2 years but older data from extensive local registries have been imported to SWEDCON as well as data from the previous GUCH registry which was started in 1992.

**Methods:** Data from SWEDCON were analyzed concerning different aspects of the transition from pediatric cardiology to GUCH.

**Results:** At the start in February 2009 SWEDCON included a total of 21 387 patients, 6815 of these came from the GUCH registry. 5491 pts were older than 19 yrs but had not been registered as GUCH patients. To our surprise this group included some patients with transposition, single ventricle, Fallot etc. Partly this could maybe be explained by follow-up at non-GUCH clinics or missed registrations but it could also be an indication that follow-up had been missed.

VSD dominated as main diagnosis in the pediatric group and ASD/PFO in the GUCH group. In the group of patients that had actively been referred from pediatric cardiology for follow-up at a GUCH clinic (1759 pts) the dominating main diagnosis was aortic or pulmonary stenosis. In the referred group 32% had a complex malformation and 72% had been operated.

All registered deaths were analyzed and the age of death was noted for different diagnoses. Early death was noted more often for complex malformations. In a few cases death as late as after the age of 60 yrs was noted even for some patients with complex disease.

From the pediatric material the number of patients between 13–18 yrs was analyzed for some diagnostic groups, where follow-up at a GUCH clinic would be mandatory. This gave a good estimate of the needed GUCH capacity during the coming 5 years.

**Conclusions:** Data from a registry such as SWEDCON can be used to improve the transition from pediatric care to GUCH care and to detect changes in referral patterns early. Long-time survival can be studied and the future need for GUCH resources can be estimated.

#### O1-4

##### **Risk stratification for congenital heart surgery in adults – Are the Aristotle Complexity score models appropriate?**

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**Introduction:** The predictive power of the Aristotle Basic Complexity (ABC) and the Aristotle Comprehensive Complexity (ACC) score for hospital mortality after congenital heart surgery has been demonstrated for children, but not for adults. We sought to evaluate the ABC, and the ACC score for adults, and to improve the discriminative power of the ACC score by adding risk factors, specifically present in adults, and not considered in the ACC score.

**Methods:** Data of all consecutive patients aged 16 or more who underwent surgery for congenital heart disease between 2005 and 2008 at our institution were collected. ABC and ACC scoring were performed according to the Aristotle Institute guidelines (Denver, USA). The impact of 5 potential risk factors for hospital mortality, not considered in the ACC score, was analyzed by logistic regression modeling. The odds ratios of significant risk factors were added to the ACC score to compile the modified ACC score. The discriminatory power of the scores was assessed using the area under the receiver operating characteristics (AuROC) curve.

**Results:** 770 procedures, performed on 495 patients during 538 operations were eligible for scoring. Hospital mortality was 2.4%. Among potential risk factors not considered in the ACC score, NYHA class >II ( $p = 0.003$ , OR = 5.4), systemic right ventricle ( $p = 0.064$ , OR = 4.4), and single ventricle ( $p = 0.001$ , OR = 8.6) were significant predictors for hospital mortality. Mean ABC, ACC, and modified ACC scores of the operations were  $6.6 \pm 2.3$ ,  $9.0 \pm 3.7$ , and  $10.6 \pm 5.6$ , respectively. The prognostic significance of the ABC and the ACC score is low when judged by the AuROC curve: AuROC of ABC score = 0.661 (95% CI 0.567–0.755,  $p = 0.047$ ); AuROC of ACC score = 0.755 (95% CI 0.668–0.841,  $p = 0.002$ ). The best discriminative power is reached by the modified ACC score: AuROC of modified ACC score = 0.852 (95% CI 0.770–0.933,  $p < 0.001$ ).

**Conclusions:** Adults requiring surgery for congenital heart defects present with additional risk factors that are not present in children. Hence, the ABC, and ACC scores are not useful for risk stratification in adults. The modified version of the ACC score discriminates well between patients with higher or lower risk for hospital mortality since risk factors, specifically present in adult patients are considered.

#### O1-5

##### **Pulmonary Arterial Hypertension in adults with Isolated Atrial Septal Defects**

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**Background:** Natural course of atrial septal defect (ASD) is complicated by pulmonary arterial hypertension (PAH). Its optimal treatment strategy is unclear.

**Aims:** To identify clinical/genetic risk factors for development of PAH in patients with ASD and to define predictors of PAH resolution and functional status after defect closure.

**Methods and Results:** 1203 patients with isolated ASD, originating from the Dutch CONCOR registry, a nationwide registry of adult patients with congenital heart defects, were included in this retrospective study. 71 patients with PAH (6%) were identified, either by echocardiography or heart catheterization. Sinus Venosus type ASD and duration of unrepaired ASD, but not gender, were predictive for the presence of PAH.

In a subsequent case-matched study of 68 ASD-PAH patients and 184 ASD-non-PAH patients, matched for ASD-type, age at defect closure and gender, no additional risk factors for PAH could be identified. 56 ASD-PAH were tested for BMPR2-mutations, no mutations were found.

A larger diameter of the ASD, older age at ASD-repair and the presence of PAH were all associated with worse NYHA functional class.

In 43 of the 68 patients with ASD-PAH, ASD closure was performed and in 27 (63%) PAH resolved after repair. No pre-operative factors (gender, NYHA class, ASD-diameter, age at repair or hemodynamics) were predictive for post-operative PAH-resolution. However, mean NYHA class improved significantly after ASD-closure, irrespective of resolution of PAH. However, mortality was higher in patients with PAH compared to those without PAH (4/68 vs. 2/186;  $p = 0.05$ )

**Conclusions:** Sinus Venosus type defect and later age at ASD-repair, but not gender or BMPR2-mutations were predictive for the development of PAH in ASD-patients. Early defect closure will reduce the prevalence of PAH in ASD-patients. ASD-repair in patients with associated PAH resulted in resolution of PAH in 63% of the patients and, independent from PAH-resolution, improved functional class. However long-term resolution of PAH remains a matter of concern.

#### O1-6

##### **Assessment of transient elastography (FibroScan®) and liver function in patients with Fontan circulation**

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**Objective:** Hemodynamic characteristics of the Fontan circulation are increased systemic venous pressure and reduced cardiac output. This altered hemodynamic status may result in chronic liver dysfunction thus leading to liver cirrhosis with the risk of hepatocellular carcinoma. Early detection of liver fibrosis is mandatory for identifying patients at risk. Transient elastography is a noninvasive, rapid method to assess liver fibrosis by measuring liver stiffness. However, there are limited data concerning the feasibility of this method in patients with a Fontan circulation.

**Methods:** A total of 24 patients after Fontan procedure with a median age of 15 yrs were studied by transient elastography using a FibroScan® (Echosens, Paris) from August 2008 to January 2011. Values were correlated with clinical (invasive hemodynamic data, echocardiography, VO<sub>2</sub>max, fenestration in conduit) and biochemical parameters (liver enzymes, BNP, coagulation tests).

**Results:** Measurements of liver stiffness could be performed in 23 of 24 patients who underwent a Fontan procedure. Liver stiffness increased directly after Fontan completion and was significantly higher compared with age-matched healthy controls ( $16.9 \pm 5.4$  kPa vs  $4.8 \pm 1.2$  kPa,  $n = 17$ ) or compared with patients with a bidirectional cavopulmonary anastomosis (BDCPA) ( $5.4 \pm 1.7$ ,  $n = 8$ ,  $p < 0.0001$ ). No significant difference was seen between patients with a BDCPA and healthy controls. Liver stiffness did not depend on the time interval since Fontan completion. There was no significant difference in liver stiffness in patients with ( $n = 10$ ) or without ( $n = 13$ ) a fenestration in the conduit. Hemodynamic parameters obtained by cardiac catheterization (mean pressure in IVC and PA as well as EDP in the ventricle) did not correlate with the data from transient elastography ( $n = 12$ ). However, liver stiffness correlated positively with BNP and GPT and negatively with ATIII ( $p < 0.05$ ).

**Conclusion:** Liver stiffness as assessed by transient elastography increases rapidly after completion of the Fontan procedure, while patients after BDCPA do not show any difference from normal controls. Biochemical parameters of impending liver dysfunction and cardiac failure in Fontan patients correspond to a further increase in liver stiffness. Therefore, transient elastography may be a sensitive diagnostic tool for early detection of patients at high risk for deteriorating liver and hemodynamic function.

#### O1-7

##### **Neoaortic root in children with transposition of the great arteries (TGA) after arterial switch operation (ASO)**

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**Objective:** Neoaortic root dilatation was described as a factor related with neoaortic valve regurgitation (NeoAR) – one of the most frequent complication after switch procedure for transposed great arteries.

This study was aimed at assessing aortic root diameters in long term observation after arterial switch. We evaluate the correlation between neoaortic root diameters or calculated indexes and neoaortic regurgitation incidence and development.

**Methods:** Among 545 patients with TGA, who had an arterial switch procedure from the year 1991 to 2009 in Cardiosurgery Department of Polish Mother's Memorial Hospital, 172 were qualified for this study. Inclusion criteria were: over 10 years of follow-up with at least 2 echocardiographic examinations in post-operative period performed in our Department with full aortic root measurements including: aortic annulus (AA), aortic sinus (AS) and sinotubular junction (STJ) diameters. Patients with two stage operation were excluded from this study.

Subjects were divided into three groups: Group 1 – patients with simple TGA (109); Group 2 – patients with TGA associated with ventricle septal defect (51); Group 3 – patients with TGA associated with aortic arch anomaly (12). Each group was divided into patients, who developed neoaortic regurgitation and those who had no signs of NeoAR during whole follow-up.



Aortic root measurements were normalized using ratio to aortic annulus (AA) and body surface area (BSA), because received values were time depending. Each year values were analyzed separately.

**Results:** Neoaortic regurgitation occurred in 85 patients (49%): 52 from group 1 (48%); 27 from group 2 (53%); and 6 from group 3 (50%). In majority of cases (84%) it was trivial/mild regurgitation, which occurred and developed in first 6 years after switch operation. None of analyzed root diameters and ratios was statistically significantly correlated with NeoAR occurrence and development.

There were no significant changes in the value of AS/AA, STJ/AA and AS/STJ indexes during whole follow up in 3 analyzed groups, while AS/BSA and STJ/BSA ratios decreased permanently with time after surgery simultaneously in three groups – BSA value increases faster than aortic root diameters during child growth.

## O2-1

### Shox2 in Pacemaker and Epicardial Development

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**Background:** The heart generates from two cardiogenic fields in the dorsal mesocardium: the first and second heart field. The first heart field gives rise to the primary heart tube and the second heart field adds cardiomyocytes to the venous as well as the arterial pole of this tube. At the venous pole these cardiomyocytes are derived from a subgroup of the second heart field, the Posterior Heart Field (PHF). Shox2 has an important role in formation of the PHF that contributes to major parts of the cardiac inflow tract including the specialized conduction system and the epicardium. We hypothesize that mutation of Shox2 leads to abnormal sinoatrial node (SAN)/pacemaking function as well as abnormal epicardial development during cardiogenesis.

**Methods:** For the assessment of embryonic heart rate and atrioventricular conduction time, electrophysiological recordings were performed in isolated hearts of wildtype and Shox2<sup>-/-</sup> embryos of 12.5 days post conception (dpc). Furthermore, immunohistochemical analysis was performed with antibodies specifically against MLC-2a, Nkx2.5, HCN4 and Wt1, and 3D-reconstructions were made.

**Results:** Compared to wildtype, Shox2<sup>-/-</sup> embryos showed a significant lower heart rate ( $105 \pm 36$  bpm vs  $74 \pm 15$  bpm;  $P = 0.032$ ), no differences were observed in atrioventricular conduction time ( $76 \pm 24$  ms vs  $80 \pm 14$  ms;  $P = ns$ ). Immunohistochemical analysis and 3D-reconstructions showed hypoplasia and aberrant differentiation of the PHF derived sinus venosus myocardium in Shox2<sup>-/-</sup> embryos of 12.5 dpc. In both wildtype and Shox2<sup>-/-</sup> embryos HCN4 was widely expressed throughout the complete sinus venosus myocardium including the SAN. The expression of Wt1 and 3D-reconstructions of the pro-epicardial organ (PEO) in Shox2<sup>-/-</sup> embryos showed a decreased PEO size at 9.5 dpc with normal epicardial spreading at 12.5 dpc. At latter stages the ventricles showed decreased numbers of epicardium derived cells and abnormal ventricular wall morphology.

**Conclusions:** Shox2 has a crucial role in venous pole development of the heart including the SAN and its pacemaking function. Furthermore, we demonstrate that Shox2 is essential for proper epicardial lineage development.

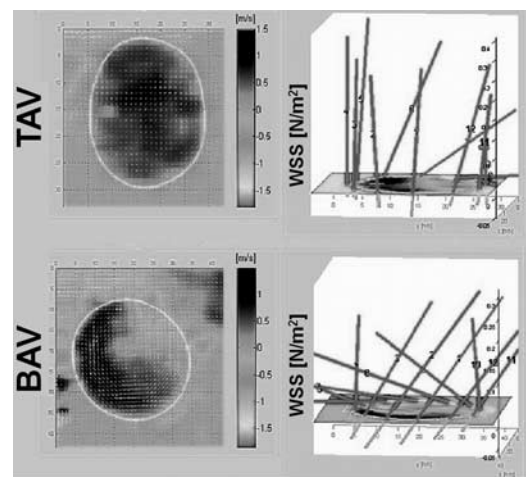
## O2-2

### Wall shear stress in the ascending aorta in patients with bicuspid aortic valves differs significantly from tricuspid aortic valves

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**Introduction:** Bicuspid aortic valves (BAV) are frequently associated with dilation of the ascending aorta. The dilation may yield in aneurysm formation and dissection of the aorta. Connective tissue defects as cystic media necrosis or dilation secondary to altered blood flow patterns in the ascending aorta are discussed to cause vessel disease in BAV. We evaluated different wall shear stress (WSS) patterns in the ascending aorta of BAV patients compared to individuals with tricuspid aortic valves (TAV) using four-dimensional cardiovascular magnetic resonance (CMR).

**Methods:** Eighteen healthy individuals with normally functioning BAV, without aortic stenosis, no aortic regurgitation, no dilation of the ascending aorta and no coarctation were compared with an age and sex matched control group of volunteers with TAV. 4D blood flow data were obtained by CMR (spatial resolution =  $2.1 \times 1.7 \times 2.5$  mm<sup>3</sup>, temporal resolution = 39.2 ms). Visualization and WSS measurement were performed with specific software tools. Measurement of the WSS in the ascending aorta was performed in the mid ascending aorta approximately on the level of the main pulmonary artery and in the proximal aortic arch at the level of the origin of the brachiocephalic trunk.



**Results:** WSS in the ascending aorta is significantly higher in patients with BAV compared to TAV. The mean magnitudinal WSS in the ascending aorta was  $0.60$  N/m<sup>2</sup> in patients with BAV and  $0.49$  N/m<sup>2</sup> in TAV ( $p = 0.028$ ). At the level of the proximal aortic arch the magnitudinal WSS did not show a significant difference between BAV and TAV. The figure shows the visualization of blood flow in the ascending aorta in two 32-year old women. The left panels show time-resolved 2D flow

profiles and the right panels show the magnitude-vector profiles of the WSS in the ascending aorta.

**Conclusions:** WSS patterns in the ascending aorta in patients with BAV without concomitant valve or vessel disease are significantly different compared to TAV. Those significantly higher shear forces may have an important impact on the development of aortic dilation in patients with BAV and are more likely to be the driving force for aortic dilatation than suggested connective tissue disorders.

## O2-3

### Array-CGH based detection of genomic imbalances in patients with heart defects as part of complex syndromes

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The prevalence of congenital heart defects (CHD) is estimated at 7–9 per 1000 births. Besides single gene mutations chromosomal aberrations are found in a significant number of syndromic and non-syndromic patients affected by CHD. In order to identify novel gene loci associated with CHD we mined clinical and experimental data from a population of patients in which we had performed array comparative genomic hybridization (aCGH) analysis for the detection of constitutional imbalances because of a variety of complex phenotypes. Array CGH had been performed using different BAC (36 K) and oligonucleotide (44 K, 105 K, 180 K, 244 K) platforms. Well-defined syndromes associated with CHD like microdeletions in 7q11 and 22q11 or Noonan syndrome were excluded. In 28 of 89 (32%) patients aCGH detected chromosomal imbalances which in part were subsequently confirmed by independent techniques like FISH or qPCR. In 8 cases the imbalance has been proven to be de novo whereas in 8 of cases the imbalance has been transmitted from one of the parents. The size of the aberrations ranged between 0.07 Mb and 151.8 Mb (median 4 Mb). Three chromosomal regions were affected in more than one patient: Two patients with discordant cardiac phenotypes had overlapping deletions in 21q22.3 (sizes 6.15 and 6.9 Mb). Overlapping deletions in 1p36.33 (sizes 3.8 and 12.7 Mb) were detected in two patients with aortic coarctation and left ventricular non-compaction cardiomyopathy, respectively. Finally, two almost completely overlapping terminal deletions in 5p15.33 distal of the Cri-du-Chat syndrome critical region of 4.2 Mb respectively 4.4 Mb were detected in two patients with a ventricular septal defect (VSD). In summary, our study showed that in nearly one third of patients presenting with complex phenotypes including CHD a constitutional imbalance can be detected by aCGH. The minimally overlapping regions affected in these patients might point to the position of candidate genes involved in the pathogenesis of CHD.

## O2-4

### Prevalence of mutations in the RAS/MAPK signalling pathway in pre-adolescent children with hypertrophic cardiomyopathy

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**Introduction:** Recent studies have shown that most cases of apparently idiopathic hypertrophic cardiomyopathy (HCM) in children are caused by mutations in cardiac sarcomere protein genes. Noonan syndrome and related malformation disorders are caused by mutations in genes encoding components of the RAS/MAPK signalling pathway and are commonly associated with HCM. Although the diagnosis is based on typical phenotypic features, in some cases the dysmorphic manifestations can be subtle. We hypothesized that mutations in the genes encoding components of the RAS/MAPK pathway cause apparently idiopathic HCM in pre-adolescent children.

**Methods and Results:** Seventy-eight patients diagnosed with HCM aged  $\leq 13$  years underwent clinical and genetic evaluation. The entire protein coding sequence of 9 RAS/MAPK pathway genes (PTPN11, HRAS, BRAF, MAP2K1 (MEK1), MAP2K2 (MEK2), KRAS, SOS1, RAF1 and NRAS), together with CBL (exons 8 and 9) and SHOC2 (4A > G mutation) were screened for mutations. Five probands (6.4%) carried novel sequence variants in BRAF, MAP2K1, MAP2K2 and SOS1 (2 individuals). Two individuals also had mutations in the MYBPC3 sarcomere protein gene.

**Conclusions:** This study identifies several novel and potentially pathogenic sequence variants in children with non-syndromic HCM. These findings have important implications for the evaluation and management of children with HCM, and provide an insight into the pathogenesis of apparently idiopathic and sarcomeric HCM.

## O2-5

### Hypothermia suppresses inflammation via NF $\kappa$ B and pSTAT3 signalling pathway in stimulated microglial cells

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**Background:** Deep Hypothermia is a standard method for neuroprotection during pediatric cardiac surgery. Today, the most serious risk factors affecting the long-term neurological outcome are neurological complications occurring during and after corrective surgery. However, the cellular mechanisms which are induced by deep hypothermia have not been clearly understood. Therefore, we investigated the effects of deep hypothermia and rewarming on BV-2 microglial cells.

**Methods:** BV-2 microglial cells are exposed to 17°C for 2 hours, slowly rewarmed to 37°C within 2 hours and observed under normothermic conditions for an additional 24 hours. For stimulation cells were treated with 1  $\mu$ g/ml Lipopolysaccharid (LPS). Cells were stained with DAPI and IB4 to detect morphological changes during cooling and rewarming. The viability of BV-2 microglial cells was quantified using a MTT assay. The pro-inflammatory cytokine IL-6 and chemokine MCP-1 secretion was measured by ELISA. The protein expressions of I $\kappa$ B $\alpha$  and pSTAT3 were analyzed using western blotting.

**Results:** Cell viability was observed to be temperature independent in the unstimulated group during the experimental period (24 hours). However, deep hypothermia led to morphological changes from a ramified and resting status under 37°C to amoeboid shaped cells under 17°C, even without LPS stimulation. The IL-6 secretion was significantly decreased 4 hours after the start of the experiment in the hypothermic group. Interestingly, after 24 hours the IL-6 secretion was equal for the hypothermic and normothermic group. Additionally, MCP-1 release was significantly decreased after 4, 6 and 24 hours under hypothermic conditions. Under hypothermia the degradation of IκBα was delayed and pSTAT3 remained down regulated at 24 hours.

**Conclusion:** Deep hypothermia had no influence on the cell viability but led to morphological changes in BV-2 microglial cells. The IL-6 and MCP-1 secretion was significantly decreased under hypothermic conditions. The regulation of the transcriptional factors pSTAT3 and IκBα were temperature dependent. Hypothermia significantly reduced the inflammatory response in stimulated microglial cells. Intervention in the inflammatory process of immunomodulatory microglial cells by hypothermia offers an interesting therapeutic option to prevent neurons from cell death.

## O2-6

### **Infection-Induced Coronary Dysfunction and Systemic Inflammation in Piglets Are Dampened in Hypercholesterolemic Milieu**

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**Introduction:** The synergism of infection with conventional cardiovascular risk factors in early life atherosclerosis is much debated. We hypothesized that coronary arterial injury correlates with infection recurrence and pathogen burden, and is further aggravated by hypercholesterolemia.

**Methods:** Forty-two Göttingen minipigs were assigned to repeated intra-tracheal inoculation of either saline, Chlamydia pneumoniae (Cpn), or both Cpn and influenza virus at 8, 11, and 14 weeks of age. Animals were fed either standard or 2% cholesterol diet (chol-diet). At 19 weeks of age coronary vasomotor responses to acetylcholine (Ach) and adenosine were assessed in vivo, and blood and tissue samples were collected. Nonparametric tests were used to compare the groups.

**Results:** In cholesterol fed animals, total cholesterol/HDL was significantly increased in infected animals ( $P = 0.01$  vs. non-infected). C-reactive protein (CRP) rose in infected animals ( $P < 0.01$  vs. non-infected) without significant difference between the mono- and co-infected groups. Among co-infected animals, both CRP and haptoglobin were lower in those fed chol-diet compared to animals fed standard diet ( $P < 0.05$ ). The vasoconstricting response to Ach was most prominent in co-infected animals ( $P = 0.03$  vs. non-infected, and  $P = 0.07$  vs. mono-infected). Among mono-infected animals, similar to CRP, a trend for less vasoconstriction was observed in those fed chol-diet ( $P = 0.08$ ).

**Conclusion:** Co-infection of piglets appears to be associated with more pronounced coronary muscarinic vasomotor dysfunction. In mono-infected animals, use of chol-diet seems to dampen both coronary dysfunction and systemic inflammation induced by infection.

## O2-7

### **Spontaneous baroreflex sensitivity in adult rats hypoxic neonatally**

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**Introduction:** We have previously shown that neonatal hypoxia is associated with an increase in systolic arterial pressure in 2 month old rats. To determine whether alterations in autonomic control are implicated in this increase we have examined baroreflex sensitivity.

**Methods:** Eighteen adult, 60 day old, Sprague-Dawley male rats were studied. Half of the animals were neonatally hypoxic (NH) ( $FiO_2 = 0.12$ , days 1–10 of life) while the others were not (Control). The rats were instrumented with intravascular telemetric blood pressure transmitter probes (Data Sciences International) to monitor abdominal aortic pressure. One week following instrumentation systemic arterial pressure was continuously monitored by telemetry for 24 hours. Systolic blood pressure (SBP) and pulse interval (PI) were determined for all beats over thirty minute periods at noon and midnight. Sequences of three or more consecutive heart beats with directionally similar changes in SBP and PI were noted. Over 5,000 sequences during nighttime and daytime in 18 rats were individually plotted as PI versus SBP. The average of all slopes with a correlation coefficient greater than 0.85 was taken as an index of baroreflex sensitivity.

**Results:** As we previously reported systemic arterial pressure was greater in 2 month old rats that had been hypoxic neonatally (Night-time: NH; systolic  $130 \pm 3$  mmHg, diastolic  $85 \pm 2$  mmHg. Control; systolic  $121 \pm 3$  mmHg, diastolic  $81 \pm 2$  mmHg; Daytime NH; systolic  $123 \pm 2$  mmHg, diastolic  $79 \pm 1$  mmHg; Control: systolic  $116 \pm 3$  mmHg, diastolic  $76 \pm 2$  mmHg,  $P < 0.05$ ). Baroreflex sensitivity was significantly less during nighttime but not daytime in the rats hypoxic neonatally (night-time: NH =  $0.84 \pm 0.09$  msec/mmHg, Control =  $1.24 \pm 0.17$  ( $P < 0.05$ ); daytime: NH =  $1.18 \pm 0.07$ , Control =  $1.43 \pm 0.14$ ).

**Conclusions:** Our results indicate that baroreflex sensitivity is decreased in adult rats hypoxic neonatally during their active night-time period. Although baroreflex sensitivity was also less during daytime this did not achieve significance. This suggests that neonatal hypoxia has longterm effects on autonomic control of systemic arterial pressure during activity. Impaired baroreflex sensitivity has been suggested to be involved in the development of systemic hypertension. This work may have important implications for the care of a growing population of adult “survivors” of congenital cyanotic heart disease.

## O3-1

### **Ebstein's anomaly and tricuspid valve dysplasia: prognosis after diagnosis in utero**

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**Introduction:** Tricuspid valve malformations are rare congenital heart diseases. The prenatal diagnosis of Ebstein's anomaly (EA)



and Tricuspid valve dysplasia (TVD) are associated with high mortality. There are conflicting reports concerning accurate prognostication after diagnosis in utero. The aim of our study was to assess prognostic factors based on our experience.

**Methods:** We retrospectively reviewed echocardiography of 37 fetuses, 26 EA and 11 TVD, between 1984 and June 2010, from 7 cardiopediatric centers. There were 11 terminations, 5 intrauterine deaths, 7 neonatal deaths and 14 survivors to age over 2 years.

**Results:** We found that the major prognostic factor for outcome was the pattern of pulmonary valve flow. Retrograde pulmonary valve flow had a predictive positive value (PPV) for death of 83%, and when associated with significant pulmonary insufficiency, PPV was 100% for death. An anatomic pulmonary atresia was also associated with 100% of death. By contrast, cardiothoracic index, right to left ventricular ratio, Celermajer index were not useful prognostic markers. Compared with retrograde pulmonary valve flow, antegrade flow in utero predicted good outcomes with a PPV for survival of 86%.

The SAS score, more complex, was less correlated to our series with 79% PPV of death when the score was  $\geq 5$  and a PPV survival of 79% when the score was  $< 5$ .

**Conclusion:** Pulmonary artery valve flow is a simple and excellent prognostic factor when major tricuspid valve disease is diagnosed in utero. Nevertheless, these fetuses should be monitored throughout the pregnancy because several hemodynamic factors may change the prognosis.

**Key Words:** Ebstein, Tricuspid valve dysplasia, Prognosis, Fetal.

### O3-2

#### **Prenatal diagnosis of complex congenital heart disease – success and outcome: A 28-year retrospective study**

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**Objectives:** Major advances have occurred in the fetal detection of, and postnatal surgery for, complex congenital heart disease (CCHD) during the past 30 years. The purpose of our study was to assess population-based changes in the detection rate and outcomes of CCHD in British Columbia (BC).

**Methods:** Between 1981 and 2008 there were a total of 7,911 pregnancies referred to our institution, the only tertiary care centre for fetal echocardiography in BC and 687 CCHD were identified. They were classified into 7 groups: hypoplastic left heart (HLH), hypoplastic right heart (HRH), univentricular heart (UNI), complex defect (CPLX), conotruncal defects (CONO), atrioventricular septal defect (AVSD), and others (OTH). The study period was divided into 7 eras of 4 years each. After diagnosis, outcomes were classified as surgical repair, termination, compassionate care, stillbirth or unknown. Survival was calculated using Kaplan–Meier analysis.

**Results:** The overall rate of prenatal diagnosis of CCHD increased constantly from 1.7/100,000 pregnancies in the first era (1981–1984) to 95.2/100,000 in the last one (2005–2008). This improvement was observed in all groups of CCHD during the first 16 to 20 years, HLH, COA, AVSD reached a plateau in their detection rate; whereas, CONO, UNI, HRH continued to show an increase in their detection rate. After the diagnosis, parents opted for termination in 294 cases (43%) and surgical repair in 241 (35%). There were 89 fetuses with chromosomal and 47 with

extracardiac abnormalities. For those opting for surgical repair, the overall 5-year survival was 77%, and there was no significant difference across eras.

**Conclusion:** Prenatal detection of CCHD has increased 50-fold during the past 28 years, reflecting major improvements in technology, knowledge, experience and training. The CONO group showed the greatest rate of diagnostic improvement. This is probably due to the inclusion of outflow tract views as part of screening protocols. The most common choice made by parents after diagnosis of CCHD was termination. Extra-cardiac and chromosomal abnormalities did not influence this decision. Survival for those having a surgical repair is good and did not differ across eras; however, more complex surgeries in the recent eras make comparisons difficult.

### O3-3

#### **Features and outcomes of cases with laterality defects diagnosed in fetal life**

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**Background:** From the postnatal data we know that congenital heart defects (CHD) associated to laterality defects or heterotaxy syndromes have a worse prognosis. Therefore, prenatal counseling is difficult in these cases. In this study we aimed to analyse the outcomes of this category of CHD diagnosed already during the fetal life.

**Material and methodology:** Out of 5800 fetuses at risk for CHD examined between 1995 and Dec. 2010 by echocardiography, 1150 had CHD and 71 (6.2%) presented an abnormal viscerotrial situs: 19 – left isomerism (Lisom), 17 – right isomerism (Risom), 12 – situs inversus with dextrocardia (SVI-dx), 4 – SVI with levocardia (SVI-levo) and 19 situs solitus with dextrocardia (SSol-dx). Anatomical features of the fetal heart, association with chromosomal or extracardiac anomalies (CA, ECA), course in utero and after birth were analysed retrospectively from the data base and clinical documentation.

**Results:** Fifty nine fetuses (83.1%) had associated CHD, in 2 with CA (trisomy 18 and deletion 6, in SVI-dx and SSol-dx) and in 3 with ECA. Among CHDs the most frequent was complex atrioventricular defect – in 29/59 (49%) cases, in Lisom, Risom, SVI-dx and SSol-dx, with atrioventricular block (AVB) in 3, followed by complex DORV (9 cases) and UVH (7 cases, 2 with AVB); 3 fetuses had corrected TGA, 3-VSD, 2 – pulmonary atresia +VSD, 2-tricuspid atresia, and one each had truncus, coarctation, partial pulmonary venous drainage and DOLV. Twelve fetuses presented isolated anomaly of the viscerotrial situs (4/12 SVI-Dx, 2/4 SVI-Levo, 5/19 SVS-dx and 1 Lisom).

**Outcome:** 18/59 cases (30.5%) opted for the termination of pregnancy, 4 died in utero (2 Lis, 1 SVI-dx, 1 SVS-dx), 15 died after birth post surgery or pacemaker implant, 2 spontaneously. Total mortality was 20/41 cases with CHD continuing pregnancy (48.8%), 56.25% (9/16) in Lisom, 45% (5/11) in Risom, 50% (5/10) in SSol-dx, 2/3 in SVI-dx and none in SVI-levo. One of 12 cases without CHD who had ECA, died after birth.

**Conclusions:** Our fetal cases with laterality defects and CHD presented a relevant mortality, mainly those with isomerisms and SSol-dx. This fact should be taken in account at prenatal counseling.

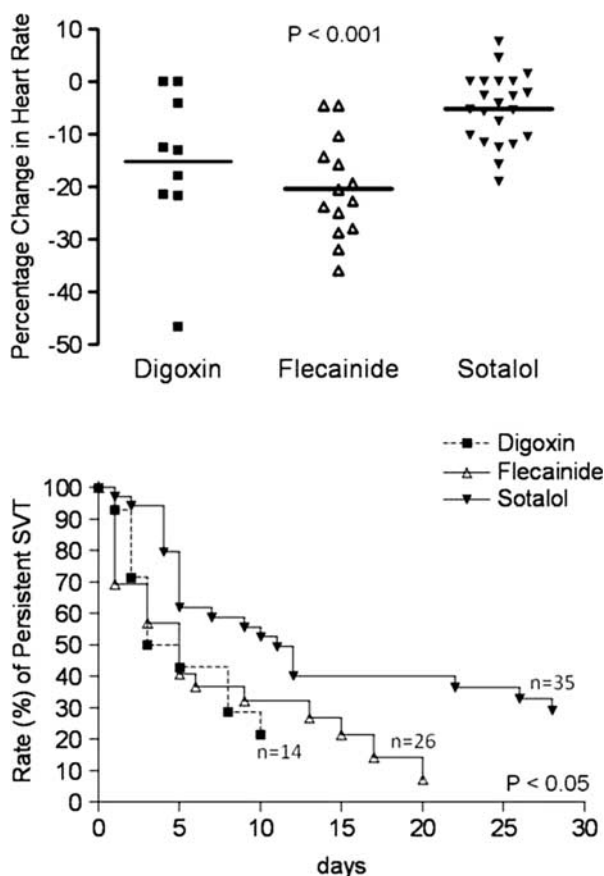
### O3-4

#### **Comparison of Transplacental Treatment of Fetal Supraventricular Tachyarrhythmia with Digoxin, Flecainide and Sotalol: Results of a Non-randomized Multicenter Study**

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**Background:** Fetal tachyarrhythmia may result in low cardiac output and death. Consequently, antiarrhythmic treatment is offered in most affected pregnancies. We compared three drugs commonly used to control atrial flutter (AF) and other forms of supraventricular tachycardia (SVT).



**Methods:** We reviewed 159 consecutive referrals to our centers with fetal SVT ( $n = 114$ ) and AF ( $n = 45$ ). Of these, 75 fetuses with SVT and 36 with AF were treated non-randomly with transplacental flecainide ( $n = 35$ ), sotalol ( $n = 52$ ), or digoxin ( $n = 24$ ) as first-line agents. Kaplan-Meier estimates were used to determine treatment effects over time to birth.

**Results:** Regardless of the choice of therapy, fetal hydrops ( $n = 33$ ) was associated with treatment failure (hazard ratio 1.8;  $p = 0.035$ ) and in-utero death (hazard ratio 4.7;  $p = 0.003$ ) and AF was more difficult to convert to sinus rhythm prior to delivery than SVT (hazard ratio 2;  $p = 0.005$ ). Cardioversion at 5 and 10 days was achieved in 50% and 63% of treated SVT cases respectively but only in 25% and 41% of treated AF cases. If incessant AF/SVT persisted to day 5 of therapy ( $n = 45$ ), median ventricular rates declined more with flecainide (-22%) and digoxin (-13%) than with sotalol (-5%;  $p < 0.001$ ) (Figure 1). Sotalol was also least likely to convert fetal SVT to a normal rhythm over time as shown in the Figure 2 ( $p < 0.05$ ). No serious drug-related adverse events were observed but arrhythmia-related mortality was 5%. Post-hemorrhagic hydrocephalus without neurological sequelae occurred in 1% (2/159 cases). None of the mothers or fetuses experienced severe treatment-related side effects.

**Conclusions:** Transplacental flecainide and digoxin were superior to sotalol in converting SVT to a normal rhythm and in slowing both AF and SVT to better tolerated ventricular rates and therefore should be considered first to treat significant fetal tachyarrhythmia.

### O3-5

#### Critical aortic stenosis at risk of hypoplastic left ventricle: has the time come for in utero fetal intervention?

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**Objectives:** Patients at risk of evolution to hypoplastic left ventricle (HPLV) could present at the time of prenatal diagnosis with a small left ventricle (LV) or with dilated LV with critical severe aortic stenosis and signs of progression towards hypoplasia. Could aortic valvuloplasty prevent evolution towards HPLV?

**Methods:** Between January 2000 and January 2011 in our tertiary center, 16 fetuses with severe aortic stenosis were considered to be at risk of HPLV: foramen oval left to right flow, ductal retrograde and monophasic mitral flow, insufficient growth of mitral and aortic annulus. Pts were divided into two groups: Group A: 11 fetuses without fetal intervention, and Group B: 5 fetuses who underwent fetal percutaneous ultrasound-guided aortic valvuloplasty between the 21st and 29th weeks of gestation.

**Results:** Group A: The 11 fetuses evolved as HPLV.

Group B: The first procedure was performed at 25 weeks of gestation, but with a LV/RV ratio of 0.70; the baby was born as HPLV. The other 4 cases were performed between 21 and 29 weeks of gestation ( $x: 23$ ) with a LV/RV ratio  $\geq 1$ ; all of them maintained LV size, but case 2 and 3 failed to recover LV contractility. In the second case parents decided to interrupt pregnancy. Case 4 and 5 showed gradual improvement of the LV contractility during the pregnancy. Last 3 cases were born biventricular and an aortic valvuloplasty was performed the 1st day of life with immediate improvement of LV contractility and a significant increase in aortic flow.

**Conclusion:** Fetal aortic valvuloplasty could offer a possibility for biventricular repair, however we draw two comments: multicenter studies should offer a large number of patients. The first trimester evolutionary study of the fetal heart may help our knowledge to detect when is the time to perform.

### O3-6

#### Postnatal left ventricular performance in prenatally treated patients with critical aortic stenosis and endocardial fibroelastosis

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Intrauterine treatment of fetuses with critical aortic stenosis (AS) and evolving hypoplastic left heart syndrome may save the left ventricle (LV) for a postnatal biventricular circulation. However, the long-term fate of these LVs with varying degrees of endocardial fibroelastosis (EFE) is unknown. The purpose of this study was to assess LV performance in surviving infants and young children, who underwent fetal aortic valvuloplasty.

**Patients and methods:** Between 12/2001 and 1/2011 29 fetal aortic valvuloplasties have been performed in 27 patients with technical



success in 19 patients (2 patients still in-utero). Ten patients went on to a biventricular circulation with 2 infant deaths. So 8 patients formed the study group with a median follow-up of 48,4 months (12–71 months). Their median fetal pre-interventional LV long-axis z-score was 0,54 (−0,99–2,1) Clinical records, echocardiographic or angiographic data were reviewed retrospectively to assess LV performance.

**Results:** 6/8 neonates had an aortic balloon dilatation, 1 neonate a surgical aortic valvotomy in the first week of life. During the follow-up period, all but 1 patient (age 4 years) needed a Ross-Konno Operation (partial EFE resection in 1 patient) at a median age of 15 days (7d–49 months). All patients are without any clinical symptoms, but still have evidence of EFE by echo or MRI and are on ACE-inhibitor therapy, 2 additionally on antiarrhythmic therapy. 1 patient needed a pacemaker due to postoperative complete AV block. LVEDD z-score was >2 in 4 patients (median 2,1; range: 0,36 to 3,0), LV SF was <28% in 3 patients (median: 30%; range 22–45%), evidence of elevated pulmonary artery pressure was present in 1 patient (1 year postop, 50% systemic pressure by echo), LAP (available in 4/8 patients) between 11 and 14 mmHg.

**Conclusions:** Children with biventricular circulation following successful in-utero aortic valvuloplasty show satisfactory, but not normal LV performance during short-term follow-up. In most cases multiple interventions and early aortic valve replacement were necessary. EFE resection at the time of surgery may be a strategy to further improve LV performance and positively impact the long-term fate of the LV.

### O3-7

#### Fetal coarctation of the aorta – can we predict who needs surgery?

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**Introduction:** Fetal coarctation of the aorta (CoA) has high false positive rates when assessed in tertiary centres. Retrospective studies indicate that quantification of arch measurements and visualisation of isthmal features such as coarctation shelf (S) and flow disturbance (F) allow separation of hypoplastic from normal arches but their ability to predict requirement for surgery in a prospective cohort remains unknown.

**Aims:** To prospectively assess the ability of isthmal Z-scores (I) and ratio of isthmus to duct (I:D) measured in three vessel and tracheal view and visualisation of S and F to identify fetuses requiring neonatal surgery for isolated coarctation.

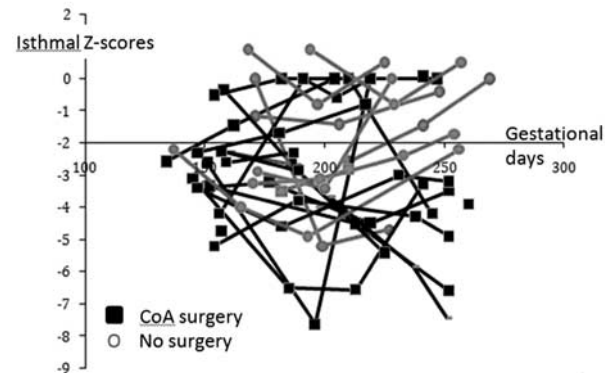
**Methods:** We acquired Doppler ultrasound measurements on 42 consecutive fetuses we diagnosed with isolated CoA referred at median 22.5 (15–37) weeks' gestation to two fetal cardiology centres. We measured I, D and I:D ratio prospectively and recorded whether S or F were seen. We recorded at the first scan and again near term whether the baby was likely to require surgery.

**Results:** 31/42 (74%) babies had CoA requiring perinatal surgery. None were diagnosed with CoA later in childhood. Both I:D ratio <0.74 (25/31, 81%) and I Z-scores <−2 (28/31 90%) gave positive predictive value for surgery of 80% at initial scan. ROC curve analysis showed a modest effect of I:D ratio (AUC 0.66, 95%CI 0.44, 0.870,  $p = 0.167$ ) and isthmal z-score (AUC 0.69, 95%CI 0.51, 0.88,  $p = 0.086$ ) to correctly identify those

requiring surgery. Serial scans were performed in 39/42 (median 3, range 1–5). Half those showing growth towards normal values (I Z-score (8) or I:D ratio (1)) or deviation from normal (I Z-score (4) or I:D ratio (4)) required CoA surgery. (figure) The variables were not powerful predictors in isolation but multi-variable logistic regression combining all gave positive predictive value 80% and ROC AUC 0.753 (0.51, 0.99).

**Conclusions:** Incorporation of these measurements in clinical practice enabled prediction of antenatal diagnosis of true CoA at first scan in 74% with correction of false positive diagnosis in 3 following serial measurements. Serial targeted examination may reduce the false positive rate of fetal CoA in clinical practice.

Figure: Serial isthmal Z scores of fetuses diagnosed with suspected coarctation



### O4-1

#### Percutaneous pulmonary valve implantation versus surgical implantation/replacement in patients with right ventricular outflow tract dysfunction

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**Objective:** Right ventricular outflow tract (RVOT) dysfunction (stenosis or regurgitation) can be treated by surgical valve implantation/replacement or by percutaneous pulmonary valve implantation (PPVI). In contrast to surgery, PPVI is usually performed in patients with dysfunction of an already existing valved conduit between RV and pulmonary artery. The aim of our study is to compare the rate of postprocedural complications, length of hospital stay and the function of the implanted valve during follow-up.

**Methods and results:** 109 consecutive patients with RVOT dysfunction were scheduled for PPVI (53) or operation (56) since December 2006. The median peak-systolic Doppler gradient across the RVOT was 69 mmHg before PPVI vs 33 mmHg before surgery ( $p < 0.0001$ ). Whereas RVOT stenosis was the prevailing lesion before PPVI, severe regurgitation was the leading lesion in the surgical group. Median age was 22.8 yrs in both groups. The median number of previous surgical procedures was significantly higher in the PPVI group (3.0 vs 2.0;  $p = 0.001$ ). The median postoperative stay was significantly longer in the surgical group (17 d vs 2 d;  $p < 0.001$ ). Post interventional complications were more frequent in the surgical group (2 patients needed urgent reoperation, 1 pacemaker insertion, 1 developed seizures, 13 fever, 4 arrhythmias) compared to those after PPVI (1 fever, 1 temporary AV-block). The median

follow-up period was identical (11.7 months). The peak-systolic Doppler gradient across the RVOT was 30 mmHg after PPVI vs 19 mmHg in the surgical group ( $p < 0.0001$ ). Substantial post-procedural pulmonary regurgitation was only present in the surgical group.

**Conclusion:** The duration of hospital stay was significantly shorter and the rate of complications after PPVI was lower than after surgery. During this time period pulmonary regurgitation was the primary indication for intervention in the surgical group whereas stenosis was the prevailing lesion before PPVI.

Therefore, PPVI seems to be the treatment of choice for selected patients with RVOT dysfunction.

#### O4-2

##### **Benefits of Stenting the Arterial Duct following Catheter Valvotomy for Pulmonary Atresia with Intact Ventricular Septum**

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**Introduction:** Catheter valvotomy is commonly accepted as the intervention of choice in neonates with pulmonary atresia with intact ventricular septum (PAIVS) and suitable anatomy. Following successful valve perforation, the greatest concern is whether the right ventricle can support the entire pulmonary flow. Therefore, in most cases, a prostaglandin infusion is continued or a Blalock-Taussig shunt (BTS) is inserted following the valve perforation. However, in our centre the arterial duct is frequently stented at the initial procedure. There are no data on the long-term outcome of this approach.

**Methods:** 37 successful valve perforations were performed between 1990–2009. The arterial duct was stented in 17 (46%) patients. Stenting was performed at the initial valvotomy procedure in 14 patients, and at a later procedure in three patients. Stenting was generally performed in those with a smaller right ventricle. Median follow-up is 8.1 years (range 1–20 years).

**Results:** Results are illustrated in table 1. There were two (13%) deaths in the stented group, one of which was partially attributed to overcirculation. There were five (25%) deaths in the unstented group. Necrotising enterocolitis (NEC) was less frequent in the stented group. One ductal stent required catheter occlusion at age four years; the remaining ductal stents occluded spontaneously. No other late complications of stenting were identified. It should be noted that the era in which the procedure was performed is a confounding factor.

Table 1. <sup>1</sup>zTV: tricuspid valve z-score (Daubeny algorithm)  
<sup>2</sup>Early reintervention: classified as reintervention <100 days post procedure

	Stented	Not Stented	P-value
Median Year of Procedure	2005 (Range 1992–2009)	1998 (Range 1990–2009)	0.003
Number of patients	17	20	
zTV <sup>1</sup>	-5.11 (±2.53)	-4.13 (±2.92)	0.283
Days ICU	4.6 (±2.9)	9.1 (±8.0)	0.029
Days in Hospital	17.4 (±18.1)	33.8 (±28.6)	0.012
NEC	2 (12%)	6 (30%)	0.246
Early Reintervention <sup>2</sup>	0	7 (35%)	0.009
Biventricular Outcome	11 (65%)	14 (70%)	0.920
Death	2 (12%)	5 (25%)	0.321

**Conclusions:** This retrospective analysis suggests that stenting of the arterial duct is an alternative to prolonged prostaglandin infusion or creation of a BTS following catheter valvotomy for PAIVS. Stenting may reduce intensive care stay and the rate of intervention.

#### O4-3

##### **Mid term outcome of arterial duct stenting: results of a multicentre study**

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**Background:** Complex CHD with duct dependent pulmonary circulation carry high morbidity and mortality in neonatal age. Although surgical palliation still represents the gold standard, percutaneous arterial duct (AD) stenting is increasingly deemed a feasible and effective alternative.

**Aim:** Aim of this multicentre retrospective study is to evaluate feasibility and mid-term outcome of percutaneous AD stenting.

**Methods:** Between March 2003 and July 2009 all the patients with duct dependent pulmonary circulation were included in the study if AD stenting was planned. Nakata and Mc Goon indexes were calculated in those who underwent a second cardiac catheterization.

**Results:** 90 patients (pts) were enrolled. Median age at the procedure was 11 days (0–502) mean weight 3.3 Kg (1.4–12.0). Diagnosis were: ToF (34.5%), PA/PS+IVS(28.5%), Ebstein TV (6%), complex CHD with PA/PS (31%). The anatomy was judged not amenable for AD stenting in 9 pts. The procedure was technically successful in 77/81 (95%). Major complications occurred in 4 pts (5%): 2 stent migration and 1 incomplete covering of the ductal tissue, requiring not urgent surgical rescue; transient cerebral ischemia in 1 successfully implanted. Operative mortality was 1,2% (1 case due to duct dissection), overall hospital mortality was 5% (4 patients: 1 due to anemia and 1 due to septicemia, 2 after Blalock Taussig shunt). At the follow-up, 5 patients(6.4%) needed a systemic-to-pulmonary artery shunt, 7 (9%) underwent percutaneous stent dilatation. Complete occlusion of the stent occurred in 1 case (1.2%), in whom PDA was the solo supply to the RPA. The Nakata and McGoon indexes calculated at follow-up angiography increased from  $132 \pm 67$  to  $287 \pm 94$  ( $p < 0,0001$ ) and from  $1,6 \pm 0,3$  to  $2,1 \pm 0,2$  ( $p < 0,0001$ ) respectively.

**Conclusion:** Stenting of the arterial duct is feasible, safe and effective palliation. It warrants an effective systemic oxygenation and promotes a significant and balanced pulmonary artery growth.

#### O4-4

##### **Wire fractures in Solysafe<sup>®</sup> Septal Occluders – a single center experience**

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**Introduction:** The Solysafe<sup>®</sup> Septal Occluder (SSO) is a relatively new device for interventional closure of patent foramina ovalia and secundum type atrial septal defects. In 08/2010, after initial unpublished reports on device fractures (DF), the manufacturer issued and “Urgent Field Safety Notice” prompting all medical care providers to reexamine all patients after implantation of a SSO (iSSO).

**Objectives:** To determine the incidence of DF after iSSO and to assess the spectrum of associated problems.

**Methods:** Prospective single center study. Extended follow-up examination including standardized fluoroscopy (sF) was performed in all patients after iSSO.

**Results:** Between 06/2005 and 07/2010, 111 patients had undergone iSSO at our institution. Median age and body weight were 50 years (9.3–79.6) and 75 kg (29–122), respectively. Indications for device implantation were 1) patent foramen ovale with a history of cryptogenic stroke (n = 84; 76%) and

2) hemodynamically significant atrial septal defect of the secundum type (n = 27; 24%). A total of 113 devices were implanted. Complete follow-up was available in 103 patients (92.8%). Median follow-up was 1.9 years (0–5.2). There were no new neurologic events or symptoms. The closure rate was 97.1%. DF was suspected by chest X-ray in one patient and documented in 10 patients by sF. In all patients with DF, damage to adjacent cardiac structures and intracardiac thrombi were ruled out by transesophageal echocardiography. The overall probability of freedom from device fracture was 82.3% after five years. There was no significant difference between the occluder sizes of 15–25 mm. The underlying cardiac lesion in all patients with DF was a patent foramen ovale (p = 0.12). In multivariate regression analysis, there was no significant influence of age, size, body weight or size at implantation. One patient had embolization of a device fragment to the right pulmonary artery. So far, all patients with DF were managed conservatively.

**Conclusions:** The incidence of DF after iSSO is acceptably high. sF is imperative for accurate diagnosis of DF. Further follow-up is needed to determine the risk for potential clinical hazards and to optimize management.

#### O4-5

##### **Premounted low profile (PLP) Stents reexpandable with over size balloons to double its original diameter. An “In vitro” evaluation study of new vascular Stents before its use in Infancy**

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**Introduction:** Although the use of intravascular stenting for vascular stenosis has been very successful and safe procedure, the application in Infancy has always been limited due to the lack of Stent adaptation to the natural growth of the vessel. Other limiting factor was to find a PLP Stent to be introduced through small size vessel adaptable for redilatation without shortening of its original length. Recently a small number of PLP stents have been designed with open struts to allow over expansion from the original radial diameter getting bigger without shortening the original length of the stent. This new concept may be applicable to small narrow vessels in infancy allowing stent redilatation to adapt the Stent diameter to the growing size of the children.

**Methods:** In Vitro evaluation of three types of vascular PLP balloon expandable Stents like:

“Formula” from Cook, “Valeo” from Edwards and “Express” from Boston Scientific.

The three types of stents had similar engineering designed with open struts and were expanded to the original diameter of the PLP balloon size of 7 mm, followed by over expansion with 14–16–18 mm balloons to double radial diameter using maximum recommended balloon pressure.

**Results:** The three types of stents when submitted to maximum balloon expansion reshaped the open struts to double the radial diameter with no shortening Stent length. When the recommended maximum balloon pressure balloon was excised the balloon exploded breaking part of the struts of the Express Stent however the other two Stents (Formula and Valeo) supported well the balloon burst keeping the new radial strength diameter. Gradual stent over expansion was also performed in aortic fresh necropsy specimen to evaluate macroscopic distension of the newborn aorta supported by over distended stent. Microscopy histological analysis of the stented aortic segment was also carried out.

**Conclusion:** PLP balloon distensible vascular Stents are now available for implant using e 6F introducer. The over distension of these new engineering designed stents allow to double the

original premounted balloon diameter. These types of stents should be considered for future used in Infancy to keep pace with somatic growth of the vessel.

#### O4-6

##### **Endovascular stenting in transverse aortic arch hypoplasia**

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**Objectives:** To describe the combined experience and outcomes from stenting transverse aortic arch hypoplasia.

**Methods:** Outcomes for transverse aortic arch hypoplasia stenting were collated from 4 centres between 2000 and 2010. Primary endpoints were reduction in instantaneous peak systolic catheter gradient, improvement in angiographic dimensions of the stented segment, and systolic right arm measured blood pressure. Changes in antihypertensive medication after stenting, early and late complications were recorded.

**Results:** 21 patients (16 male, 5 female) were included. Median age was 16.5 years (0.25–25.9 years). Median weight was 55 kg (4.5–103 kg) and median height 162.5 cm (54–182 cm). 19 patients were hypertensive at baseline. There were only 2 neonates, both post repair of interruption where there was no other surgical option for the recoarctation.

Median diameter of the native transverse arch was 7 mm, increasing to 14 mm post-stenting (p < 0.0001). Median ratio of the transverse arch to descending aorta at the diaphragm level improved from 0.43 to 0.9 (p < 0.001). Mean baseline gradient across the hypoplastic transverse arch was 38.1 mmHg (14–76 mmHg). Mean post-stent gradient was 4.95 mmHg (0–13 mmHg) p < 0.0001.

There were early complications in 5 patients (1 stroke, 2 stent migrations, 1 puncture site bleeding requiring transfusion, and 1 transient arm neuropraxia), with no deaths.

**Follow up:** Follow up data was available for 19 patients; median follow up period 24 months (1–120 months). 15 patients had a follow up CT, and 3 had an MRI. One neonate had a stent fracture and intimal hyperplasia within the stent at 12 months. A second neonate had intimal hyperplasia of the stent requiring balloon dilation of the stent at 30 months.

17/19 patients had medium term follow-up data on non-invasive blood pressure and medication. Pre-stent median systolic blood pressure was 153 mmHg (117–180 mmHg), with a post-stent systolic median of 130 mmHg (105–150 mmHg) p = 0.0002. The baseline median diastolic blood pressure was 78 mmHg (49–107 mmHg), reducing to 70 mmHg (50–90 mmHg) post-stent (p = 0.006). 13/17 patients showed a reduction in antihypertensive medication post-stent.

**Conclusions:** Stenting of transverse arch hypoplasia although technically challenging, produced good angiographic and haemodynamic results with an early improvement in blood pressure control. These results appear to be sustained in the medium term.

#### O4-7

##### **Non-invasive Cardiac Output Monitoring during Catheter Interventions in Patients with Cavo-pulmonary Circulations**

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**Introduction:** Electrical velocimetry uses changes in thoracic electrical bio-impedance to calculate cardiac output non-invasively. Bioimpedance is altered with systolic acceleration of erythrocytes in the aorta and can be used to calculate several indices related to cardiac output. Recent studies have favourably compared electrical velocimetry to previously established invasive measurements of cardiac output including thermodilution and transoesophageal Doppler. Electrical velocimetry has also been shown to be comparable to cardiac output measured using the Fick principle in children with complex congenital heart disease.

**Methods:** We used an Icon<sup>®</sup> monitor (Osypka, Germany) to assess changes in CO during catheter interventions in ten patients with cavo-pulmonary connections. Nine patients had hypoplastic left heart syndrome post fenestrated Fontan and one had pulmonary atresia, intact ventricular septum and Glenn shunt (1.5 ventricle repair). Stroke volume was recorded during periods of stable heart rate before and after interventions and so was directly related to CO. Mean patient age was 6.1 (4.8–15.3) years and mean weight was 18.5 (15–63) kilograms.

**Results:** Stroke volume and therefore cardiac output were increased in patients following stenting the left pulmonary artery (mean 16% increase). Partial occlusion and complete closure of right to left atrial shunts showed a decrease in stroke volume which may reflect a reduction in ventricular preload (mean 12% decrease). Creation of Fontan stent fenestration resulted in a marked increase in stroke volume (22% and 29% increase) in these two patients.

**Conclusions:** Icon<sup>®</sup> is a novel monitoring technique ideally suited for use during interventional catheter procedures. Initial experience in patients with Glenn shunt and Fontan circulations is promising and provides new insights into the pathophysiology of the circulation in series.

## O5-1

### Perioperative Uncontrolled Pulmonary Hypertension Alters Long-Term Function And Durability Of Right-Sided Homografts Implanted In Ross And Non-Ross Patients

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**Objectives:** To date the potential role of pulmonary hypertension on outcomes of homografts used for right ventricular outflow tract (RVOT) reconstruction has not been evaluated. The objective of this study was to prove the deleterious role of both preoperative and postoperative pulmonary hypertension on long-term function and durability of homografts implanted in pulmonary position through a multivariate risk analysis in both Ross and non-Ross patients.

**Methods:** A retrospective study (1993–2009) included 222 consecutive patients who received homografts for RVOT reconstruction. Right ventricle (RV) and pulmonary artery (PA) pressures were measured by echocardiography before and after implantation with a mean follow-up of 5.3 years (6d.–17.9 yrs). Median age at implantation was 12.5 years (4d.–67 yrs). Main heart defects were aortic valve diseases (requiring a Ross procedure) (n = 107, 48,6%), pulmonary atresia with VSD (n = 40, 18%), truncus arteriosus (n = 28, 13%). End-points were homograft stenosis (maximal echographic peak gradient  $\geq 20$  mmHg), regurgitation (grade at least moderate), dysfunction (maximal echographic peak gradient  $\geq 50$  mmHg or regurgitation at

least moderate) and explantation. Univariate and multivariate Cox regression analyses were performed.

**Results:** Univariate risk analysis is presented in the table. The multivariate risk analysis confirmed that the preoperative mean pulmonary artery pressure  $> 30$  mmHg is a significant risk factor for homograft stenosis (Hazard Ratio HR:5; p = 0,01), regurgitation (HR:3; p = 0,02) and explantation (HR:80; p = 0,03). A RV/LV pressure ratio  $> 0,5$  at 8 months after implantation without any significant homograft stenosis is a multivariate risk factor for homograft dysfunction (HR:9; p = 0,002).

**Conclusions:** Preoperative and postoperative pulmonary hypertension alters long-term function and durability of homografts implanted in pulmonary position both in Ross and non-Ross patients. A better control of the perioperative pulmonary artery pressure through surgical and medical measures could improve their durability.

Univariate risk analysis for HG-related events	Homograft stenosis	Homograft regurgitation
Preoperative risk factors	RV/LV pressure ratio $> 0,4$ (p = 0,001) RV pressure $> 40$ mmHg (p = 0,005)	Mean PAP $> 30$ mmHg (p = 0,002) RV/LV pressure ratio $> 0,4$ (p = 0,08) RV pressure $> 40$ mmHg (p = 0,008)
Immediate postoperative risk factors	Mean PAP $> 30$ mmHg (p = 0,02) PA branch stenosis (p = 0,03)	Pulmonary hypertension crisis (p = 0,01)
risk factors at 8 months after implantation	Peak PAP $> 49$ mmHg (p = 0,03) RV/LV pressure ratio $> 0,5$ (p $< 10^{-3}$ )	Peak PAP $> 49$ mmHg (p = 0,01) Distal stenosis of pulmonary arteries (p = 0,005)
Univariate risk analysis for HG-related events	Homograft dysfunction	Homograft explantation
Preoperative risk factors	Mean PAP $> 30$ mmHg (p = 0,005) RV pressure $> 40$ mmHg (p = 0,01)	Mean PAP $> 30$ mmHg (p = 0,002) RV/LV pressure ratio $> 0,4$ (p = 0,002) Distal stenosis of pulmonary arteries
Immediate postoperative risk factors	RV pressure $> 45$ mmHg at the end of CPB (p = 0,01)	
risk factors at 8 months after implantation	Distal stenosis of pulmonary arteries (p = 0,02) RV/LV pressure ratio $> 0,5$ (p $< 10^{-3}$ ) Mean PAP $> 30$ mmHg (p = 0,04)	Mean PAP $> 30$ mmHg (p = 0,001) RV/LV pressure ratio $> 0,5$ (p = 0,02)

## O5-2

### Patient age influences neo-aortic root dimensions and aortic regurgitation following the Ross operation in children

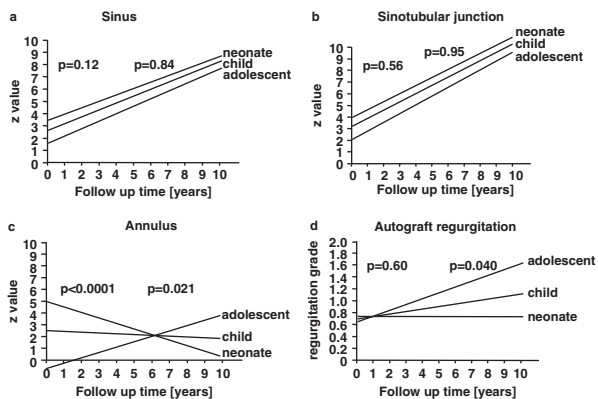
Höer J., Kasnar-Samprec J. (1), Stierle U. (2), Bogers A.J.J.C. (3), Hemmer W. (4), Hetzer R. (5), Sievers H.H. (2), Lange R. (1)  
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**Introduction:** For children who require aortic valve replacement, the Ross operation provides the advantage of growth potential of the pulmonary autograft in the aortic position. However, development of autograft dilatation and regurgitation may occur

in some patients. We sought to assess the progression of autograft diameters and regurgitation with regard to patient age.

**Methods:** Autograft echo dimensions from 48 children <16 years of age at the time of the Ross operation, who had follow-up echocardiograms at <20 years of age, were analyzed using hierarchical multilevel modeling. Z-values of autograft dimensions were calculated according to the normal aortic dimensions. Mean follow-up was  $5.1 \pm 3.3$  years.

**Results:** The mean age at the time of the Ross operation was  $10.0 \pm 4.3$  years. The mean z-values showed a significant increase with follow-up time at the sinus ( $0.5 \pm 0.1$ /year,  $p < 0.001$ ), and the sinotubular junction ( $0.7 \pm 0.2$ /year,  $p < 0.001$ ), but not at the annulus ( $0.1 \pm 0.1$ /year,  $p = 0.59$ ). There was no significant difference of the sinus, and the sinotubular junction z-value between younger and older children (figure a and b). The annulus z-value was significantly larger in younger children ( $p < 0.0001$ ), whereas the annual increase was significantly higher in older children ( $p = 0.021$ , figure c). Autograft regurgitation develops significantly faster in older children ( $p = 0.040$ , figure d).



**Conclusions:** Sinus and STJ dilate with time regardless of patient age. Young children show larger initial annulus sizes than older children. However, annulus diameters tend to normalize in young children, whereas they increase in older children. Autograft regurgitation develops in older children, but not in young children. Stabilizing measures to prevent autograft root dilatation are warranted in adolescents, but they are not required in young children.

### O5-3

#### Right Ventricular Outflow Reconstruction With Injectible Valve

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**Introduction:** Significant pulmonary regurgitation is a common problem after surgical or percutaneous treatment of congenital cardiac defects like Tetralogy of Fallot or Pulmonary Stenosis negatively affecting long-term prognosis and necessitating re-interventions.

The Biointegral no-react injectable pulmonic (NRIP) valve allows pulmonary valve replacement with or without cardiopulmonary bypass under direct control minimizing the impact of surgery on cardiac function. The aim of the works is to describe our multi institutional experience with the clinical use of this device.

**Methods:** Between January 2006 and January 2011, 21 symptomatic patients, mean age  $21.0 \pm 12.5$  (5.8–53.5), with severe pulmonary regurgitation and progressive right ventricular dysfunction after Tetralogy or similar pathology repair, received NRIP in 7 European different Institutions. All patients underwent magnetic resonance (MR) studies before and after the implant and trans-esophageal 2-D echocardiography during the surgical procedure and the follow up.

**Results:** Valve insertion, delivery, and placement were successful in all patients but one that required the repositioning of the same valve in cardiopulmonary by pass. Of these, 4 patients implanted the NRIP in cardiopulmonary by pass to allow repair of intracardiac defects. Early recovery was uneventful and all the patients were discharged home after a mean length of hospital stay of  $6.3 \pm 2.4$  (2–12) days. Intraoperatively, transesophageal echocardiography was the unique tool to guide device positioning and verify early surgical results. In two patients echo documented a valve displacement after delivery and guided the repositioning (one in cardiopulmonary by-pass). In the immediate postoperative course serial echocardiographic examinations and MR studies documented right ventricle reverse remodeling and excluded later complications. The mean follow up was 2.8 yrs (2 months – 4 yrs).

**Conclusions:** The NRIP valve allows safe and easy pulmonary valve replacement without cardiopulmonary by pass in selected cases. Its mode of implantation offer less invasive approach with less blood loss and shorter hospital stays. Trans esophageal echocardiography plays an important role in the intraoperative management and the MR permit an adequate selection of the patients. Longer follow up is required to assess the valve performance.

### O5-4

#### When Coronary Arteries Need Right Ventricular Systolic Pressure

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**Introduction:** Patients with pulmonary atresia and intact ventricular septum (PA IVS) can have coronary sinusoids connected to a hypertensive RV. Coronary perfusion can be dependent on the RV systolic pressure wave (RVDCC), even when dual supply exists. Decompression of RV then can be deleterious. We evaluated the outcome of different treatment strategies.

**Patients and Methods:** National, multicentre (4), retrospective analysis. Of 207 patients (pts) born 1985–2010 with PA IVS, 41 had coronary sinusoids. All angiograms, cardiac ultrasounds, surgical reports, pathology reports and outcome data were reviewed. **Results:** 13 patients had normal coronary flow, 17 had dual flow of which 7 had significant systolic RV flow. 11 patients had segments with only sinusoidal perfusion. Thirty five pts (85%) received a BT-shunt at 11 days (2d–16m); 22pts (53%) had a Glenn shunt at 8 months (2m–5y). Twelve pts (29%) went to full Fontan at 3y (2.5–19y).

Eleven pts (27%) had fulguration and balloon dilation of pulmonary valve, 3 with RVDCC; 2pts (5%) RVOT surgery. Only one patient with RV decompression (dual flow) died at the age of 7 months short after bidirectional Glenn due to collaps of the right coronary

artery; in retrospect RVDCC. In 3pts (7%) with dual flow, decompression resulted in disappearance of sinusoids.

Eighteen patients (43%) had RVDCC. Twelve patients died (29%), 8 (19%) with RVDCC at age 93.5 days (9d–2.5 y): 7 at BT-shunt, 2 at bidirectional Glenn; all suffered massive ischemia. 3 patients died >1y suddenly (no pathology). Pathology in 4 showed marked coronary hypertensive vasculopathy, intimal fibrosis and medial hypertrophy.

**Conclusion:** Decompression of the RV appears to be safe if dual flow without RVDCC; frequently regression of sinusoidal flow was observed. If RVDCC, reduction of coronary perfusion to “normal aortic diastolic flow” may result in acute ischemia and infarction. Cardioplegia (administration and washout) is different in RVDCC and should be avoided. Natural history of RVDCC is dreadful with progressive endothelial proliferation resulting in late ventricular ischemia. Heart transplantation is probably the best long term option in RVDCC.

#### O5-5

##### “Hybrid” palliation of Interrupted Aortic Arch as initial treatment modality

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**Introduction:** Complex interrupted aortic arch (IAA) requiring early surgical treatment can be challenging. We report our experience with the “hybrid” approach for palliation of these infants prior to elective definitive surgery to facilitate concomitant treatment of the associated lesions and to decrease complications.

**Methods:** Retrospective review of all consecutive patients with IAA that were treated by the “hybrid” approach at a single institution. Complex IAA defined as, weight of less than 2.5 kg, multiple VSD, multi-organ system failure and narrow left ventricular outflow tract. All patients were palliated in the “hybrid” catheterization laboratory by a median sternotomy, bilateral branch pulmonary arterial band to 3.0 to 3.5 mm and ductal stenting with nitinol self-expanding stent through a sheath placed directly into the main pulmonary artery. Definitive surgical repair was performed at a mean of 4.4 months. Left heart structural dimensions were measured pre-palliation and prior to surgical repair and are reported.

**Results:** Seven patients were treated from July 2007 till December 2010. Weight range 2.0 to 3.4 (mean of 2.6) kg. Associated complexity; weight less than 2.5 kg (n = 2), multiple VSD (n = 2), shock with multi-organ failure (n = 1). All “hybrid” palliations were successfully performed with no mortality. All patients were discharged home. One patient died 2 weeks post palliation from necrotizing enterocolitis. All other patients underwent successful biventricular surgical repair. One patient underwent trans-catheter device closure of a muscular VSD and another per-ventricular device closure at the time of surgical repair. Somatic growth and growth of the left heart structures are shown in Table 1.

	Mean Pre-hybrid	Mean Pre-Definitive repair	p Value
Weight (Kg)	2.6	4.7	p = 0.0007
BSA (m <sup>2</sup> )	0.17	0.26	p = 0.0006
LV Volume (ml)	2.9	4.5	p = 0.002
LV Volume z-score	-2.54	-2.84	p = 0.006
Aortic Valve Annulus (mm)	4.63	5.92	p = 0.005
Aortic Valve Annulus z-score	-2.64	-2.75	p = 0.5
Mitral Valve Annulus (mm)	9.53	11.40	p = 0.025
Mitral Valve Annulus z-score	-0.14	-0.39	p = 0.35

**Conclusions:** “Hybrid” palliation of IAA by bilateral branch PA banding and ductal stenting is feasible and provides adequate

palliation with excellent somatic growth even in those patients with additional complexities. Although LV structures did not “grow” relative to somatic growth, final repair is facilitated especially in the presence of additional VSDs. This approach provides a safe alternative strategy in complex patients with IAA.

#### O5-6

##### RV morphology remains a risk factor for early postoperative outcome after Fontan operation

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**Introduction:** We sought to assess the impact of right (RV) or left (LV) systemic ventricular morphology on early postoperative outcome after extracardiac Fontan operation (ECFO).

**Methods:** 136 consecutive patients with median age of 3,8 years and weight of 14,3 kg, Nakata index of 231, lower lobe index of 143 and mPAP of 10 mmHg underwent ECFO between 1995 and 2010. Intraoperative and early postoperative course was compared between two groups with LV (n = 87, 64%) and RV (n = 49, 36%) for the following outcomes: cardiopulmonary bypass time, cardioplegia time, early mortality, mechanical circulatory support (MCS), intensive care unit (ICU) and hospital stay. Hemodynamics were compared regarding circulatory (mean pulmonary (mPAP), left atrial (LA) and mean arterial (MAP) pressure and catecholamines requirements), pulmonary (mechanical ventilation and nitric oxide (NO) requirement) and renal failure (ascites, diuretic therapy, dialysis).

**Results:** There were no differences in pre- and intraoperative data or the need for postoperative MCS between patients with RV or LV. Early postoperative mortality was higher in RV patients (14% RV, 2% LV, p < 0.05).

RV patients displayed a higher LA pressure (9 (4–22) RV vs 6 (–1–14) LV mmHg, p < 0.05), lower MAP (55 (18–99) RV vs 59 (35–90) LV mmHg), p < 0.05) and more catecholamines requirements longer than 72 h after FO (38% RV, 14% LV, p < 0.05).

Longer mechanical ventilation (75 (1–858) RV vs 13 (2–518) LV hours, p < 0.05) and greater requirement of NO inhalation (53% RV, 29% LV, p < 0.05) was present in RV patients, but no difference in mPAP was seen between RV and LV patients. The incidence of ascites (60% RV, 30% LV, p < 0.05), requirement of intensified diuretic therapy (39% RV, 15% LV, p < 0.05) and the need for dialysis (55% RV, 5% LV, p < 0.001) was higher in RV patients.

Patients with systemic RV had longer ICU and hospital stay (6 (1–37) RV vs 3 (1–77) LV days; 18 (9–44) RV vs 14 (2–107) LV days, p < 0.05).

**Conclusion:** A systemic RV is still a risk factor for early postoperative severe morbidity. For optimal outcome in patients with systemic RV rigorous preoperative selection criteria and aggressive postoperative management are necessary.

#### O5-7

##### Levosimendan versus Milrinone after Corrective Open-Heart Surgery in Neonates and Infants

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**Introduction:** Levosimendan has been shown to improve cardiac function and hemodynamics in adults. After open-heart surgery in neonates and infants low cardiac output syndrome (LCOS)



commonly complicates the postoperative course and is associated with poor outcome. Therefore, milrinone is widely used prophylactically to prevent LCOS after open-heart surgery. The aim of our study was to evaluate whether levosimendan is superior to milrinone in preserving cardiac output (CO) after open-heart surgery in infants.

**Methods:** After written informed consent forty children <1 yr old (71 ± 80 days, 4.2 ± 1.3 kg) undergoing corrective open-heart surgery (basic Aristotle score 8.9 ± 1.8) were included in a prospective single-center, double-blind, randomized pilot-study. Exclusion criteria were <36 weeks of gestation, <3 kg weight, preoperative LCOS, pretreatment with the study drugs, renal impairment and thrombocytopenia. At weaning from cardiopulmonary bypass patients received either a 24 hours continuous infusion of 0.1 microg/kg/min of levosimendan (n = 20) or a 24 hours continuous infusion of 0.5 microg/kg/min of milrinone (n = 20). The primary study endpoints cardiac output (CO) and index (CI) using transesophageal Doppler technique (Cardio-QP, Deltex Medical), hemodynamic parameters and FS were evaluated at 2, 6, 12, 18, 24 and 48 hours post cardiopulmonary bypass. ANOVA was used for statistics.

**Results:** There were no differences in demographic data, complexity of cardiac surgery, bypass time and aortic cross clamp time. Both drugs were well tolerated and no death or serious adverse event occurred throughout the study. The duration of mechanical ventilation, stays in ICU and total hospital stay did not differ between the groups. Heart rate, systemic arterial pressure, pulmonary artery pressure, left atrial pressure, arterial to venous saturation difference, NIRS, FS, lactate levels, total volume requirement, urine output and inotrope score were similar in both groups.

In the levosimendan group compared to the milrinone group there was a statistically significant increase of cardiac output (p = 0.043) over time. However, the increase in cardiac index between the groups only showed a trend (p = 0.077).

**Conclusions:** Levosimendan was found to be safe when prophylactically given to neonates and infants following open-heart surgery. The prophylactic use of levosimendan slightly increased CI. However, this marginal hemodynamic benefit did not observably influence patients' outcome.

#### O6-1

##### 4-dimensional flow patterns in the ascending aorta differ strongly between bicuspid and tricuspid aortic valves

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**Introduction:** Bicuspid aortic valves (BAV) are frequently associated with dilation, aneurysm and dissection of the ascending aorta. Two opposing hypotheses advocate either an inborn connective tissue defect or dilation secondary to altered blood flow conditions in the ascending aorta.

This study was initiated to evaluate flow patterns in the ascending aorta of BAV patients compared to those in individuals with tricuspid aortic valve (TAV) using four-dimensional cardiovascular magnetic resonance (CMR).

**Methods:** 18 healthy individuals with normally functioning BAV, without aortic stenosis, aortic regurgitation or dilation were compared with an age and sex matched control group of volunteers with tricuspid aortic valve (TAV). 4D blood flow data were obtained by CMR (spatial resolution = 2.1 × 1.7 × 2.5 mm<sup>3</sup>,

temporal resolution = 39.2 ms) and visualization was performed with dedicated software. Evaluation of different flow patterns was performed by three blinded observers and flow alterations were classified into four groups concerning their intensity.

**Results:** In 90% BAV and TAV were correctly classified in blinded evaluation of flow visualization. Abnormal helical flow patterns in the ascending aorta were seen in 85% of the evaluations in the BAV group. In the TAV group altered flow was only found in 6%. Comparison of flow patterns in the matched pairs revealed a significant difference between patients with BAV and the control group (p = 0.0004). Figure 1.

**Conclusions:** Patients with BAV without concomitant valve or vessel disease have a significantly different 4D flow pattern than patients with TAV. This altered flow may have an important impact on the development of aortic dilation in patients with BAV.

We further suggest that connective tissue defect may only be a secondary finding in these patients.

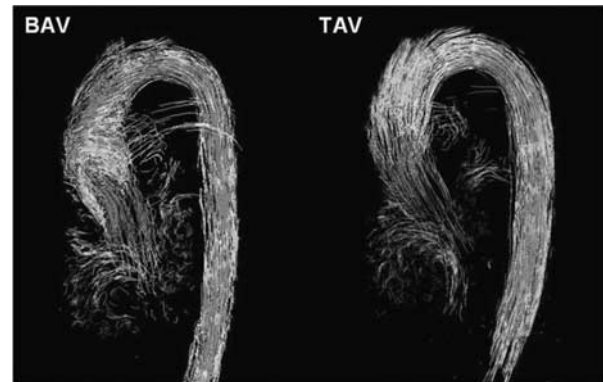


Figure 1: 4D flow visualization of blood flow of the thoracic aorta in two 32-year-old women. (BAV: bicuspid aortic valve; TAV: tricuspid aortic valve)

#### O6-2

##### The cardio renal anemia syndrome in adult patients with congenital heart disease

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**Background and aims:** Increasing numbers of reports have documented recently the close linkage between congestive heart failure (CHF) and chronic kidney disease (CKD). Both conditions can cause and worsen each other. Since these patients are often anemic, some investigators have called this vicious circle "cardio renal anemia syndrome". This study aimed to elucidate the linkage between CKD and anemia in adult patients with congenital heart disease (ACHD), and to investigate whether these factors can predict cardiovascular events.

**Methods and Results:** A total of 268 ACHD patients were enrolled (mean age 30 ± 11 years, 45% female). During a follow-up of 33 ± 13 months, 69 patients (26%) experienced a cardiovascular event (cardiac death, symptomatic arrhythmia, hospitalization because of worsening heart failure, and/or thromboembolism). The mean eGFR in these 268 patients was 83 ± 25 ml/min/1.73 sq m and the mean hemoglobin level was 15.3 ± 5.4 g/dl. The overall prevalence of CKD (eGFR < 60 ml/min/1.73 sq m) was 12%, and the prevalence of anemia (hemoglobin < 12g/dl) was 7% in this cohort. The prevalence of CKD and anemia were higher in patients with a New York Heart Association functional classification of class I or greater (p < 0.05). ACHD patients with CKD had lower hemoglobin levels and higher BNP levels compared with those of patients without CKD (p < 0.01). eGFR was related to the hemoglobin level (R = 0.39, p < 0.05), and

inversely related to the serum BNP level ( $R = 0.54$ ,  $p < 0.01$ ). Application of Kaplan–Meier analysis to the overall patient group indicated that eGFR was a significant predictor of cardiovascular events, while the hemoglobin level was not. In patients without cyanosis, both the eGFR and the hemoglobin level were strong predictors of cardiovascular events.

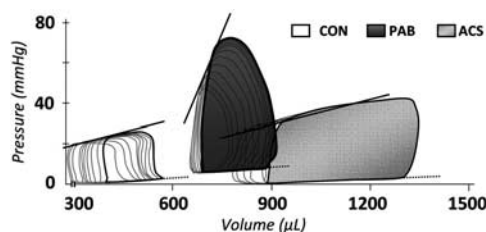
**Conclusion:** The eGFR and hemoglobin levels were closely related to each other, and both levels declined with worsening CHF. Although the hemoglobin level had a predictive value only in patients without cyanosis, eGFR was a significant predictor of cardiovascular events in the ACHD study group overall.

### O6-3

#### Distinct patterns of functional right ventricular adaptation to experimental right ventricular pressure vs volume overload

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**Introduction:** Right ventricular failure due to chronic abnormal loading conditions is a pivotal determinant of prognosis in congenital heart disease. However very little is known about the functional and biomolecular response of the right ventricle (RV) to abnormal pressure- or volume loading and consequently RV-specific treatment is lacking. Our objective was to define the RV response to abnormal loading in terms of systolic and diastolic function and hypertrophy development.



	CON	PAB	ACS
RV wt/bodywt (mg/g)	0.6 ± 0.01	1.2 ± 0.1*	1.1 ± 0.1*
PAB gradient (mmHg)	4 ± 1	76 ± 22*†	7 ± 1
Cardiac output (mL/min)	82 ± 2	70 ± 4*	126 ± 8*§
Strokework (mmHg mL)	5.2 ± 0.3	15.1 ± 1.8*	13.0 ± 1.0*
Systolic parameters			
ES elastance (mmHg/mL)	59 ± 8	155 ± 27*†	41 ± 4
Ejection Fraction (%)	47 ± 3	27 ± 2*	34 ± 2*
Diastolic parameters			
ED elastance (mmHg/mL)	4 ± 1	9 ± 2*	4 ± 2
Tau (ms)	16 ± 1	21 ± 1*†	16 ± 1
ES Volume (µL)	320 ± 42	648 ± 29*†	839 ± 49*
mRNA/18S expression			
NPPB	1.0 ± 0.3	22.3 ± 6.0*	16.2 ± 4.2*
MCIP	1.0 ± 0.2	7.0 ± 1.2*	5.1 ± 0.9*
bMHC_aMHC_ratio	1.0 ± 0.4	10.9 ± 3.2*	10.3 ± 5.6*

n = 8–10/group; \*  $p < 0.05$  vs CON, † vs ACS, § vs PAB

ES = endsystolic, ED = enddiastolic, NPPA/B = natriuretic propeptide A/B, MCIP = modulatory calcineurin-interacting protein, b/aMHC = beta/alpha myosin heavy chain

**Methods:** Wistar rats were subjected to either chronic RV pressure load by pulmonary artery banding (PAB), or chronic RV volume load by aorto-caval shunt (ACS). After 4 weeks, RV pressure-volume measurements were performed. Data are presented as mean ± SEM.

**Results:** PAB resulted in marked pressure load (banding gradient  $76 \pm 22$  mmHg) and ACS in marked volume load: cardiac output increased by 54%. All data in the table, see the figure for pressure-volume loops. Though different in nature, the absolute severity of loading appeared equal in both models (Strokework PAB vs ACS = ns). Also, hypertrophy development was equal in PAB and ACS. The classic key genes of hypertrophy regulation (NPPA, NPPB, MCIP) were upregulated in both models, but expression tended to be higher in PAB. The same was true for beta-to-alpha MHC isoform-switch. The haemodynamic responses however were very distinct: Pressure loading led to increased contractility as measured by endsystolic elastance, but insufficient to maintain cardiac output and ejection fraction. Additionally, diastolic function was impaired as indicated by enddiastolic elastance and tau and there was marked dilatation (endsystolic volume). Volume loading resulted in no changes in endsystolic elastance. Diastolic parameters were unchanged whereas the ventricles were significantly dilated.

**Conclusions:** In this study of experimental RV pressure loading vs volume loading, we show that equal magnitude of loading leads to very different functional responses, which can not be explained by changes in the classic 'LV-hypertrophy-genes'. Further analysis of these models may allow for identification of new RV-hypertrophy-genes, which pose potential targets for RV-specific therapy.

### O6-4

#### MicroRNAs may control mRNAs in circulating peripheral blood mononuclear cells during the acute phase of Kawasaki disease

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**Introduction:** MicroRNAs (miRs) are small noncoding RNAs of 18–25 nucleotides that mediate gene silencing through imperfect hybridization to 3' untranslated regions in target mRNAs. The human genome encodes more than 800 different miRs that modulate a variety of biological activities including immunological reactions. Kawasaki disease (KD) is the most common systemic vasculitis syndrome primarily affecting small and medium-sized arteries, particularly the coronary arteries. Patients are diagnosed with KD if they had 5 or more of the following diagnostic criteria: more than 5 days-lasting fever, conjunctivitis, skin rash, red palms and soles or peeling, and swollen lymph nodes. It was reported that peripheral blood mononuclear cells (PBMCs)-derived vascular endothelial growth factor (VEGF) may contribute to later vascular injury and remodeling in KD, but the etiology of KD is not clear yet. Though KD may be associated with immunological problems, the involvement of miRs in KD has not been reported.

**Methods:** We performed mRNA and miR microarray analysis of PBMCs isolated from acute KD patients (N = 4) who were responsive to intravenous immunoglobulin treatment (2 g/kg/24hr), a febrile disease group (viral or bacterial infection, N = 6), or healthy controls (N = 6). The data were analyzed using GeneSpring GX 11.0 software and Ingenuity Pathways Analysis tools.

**Results:** Prior to treatment miR-93 and miR-877 were down-regulated and miR-92b, miR-182 and miR-296-5p were up-regulated by comparison with the febrile group and healthy controls. The levels of these 5 miRs normalized after treatment. The expression of *VEGFA*, which were previously reported to be controlled by miR-93, was up-regulated prior to treatment and

normalized upon treatment, negatively correlating with the expression of miR-93.

**Conclusions:** We identified 5 miRs, miR-93, miR-877, miR-92b, miR-182, miR-296-5p, which were highly specific to the acute phase of Kawasaki disease suggesting a small number of known miRs play an important role during the acute phase of KD. In particular miR-93 may control the expression of *VEGFA* and have an important role in signaling resulting in the development of coronary artery lesions. We are currently analysing the biological function of the other 4 miRs.

#### O6-5

##### Clinical application of a vagal hyperreactive animal model

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Vagal hyperreactivity is known as a cause of vasovagal syncope, and has been proposed as a possible cause of sudden infant death syndrome (SIDS).

In a first study, rabbits with marked vagal pauses following injection of phenylephrine were selected and crossed to obtain a vagal hyperreactive (VHR) strain. In binding experiments, we observed an overexpression of M2 and M3 muscarinic receptors density in the heart of this VHR rabbit model (Bmax (fmolmg<sup>-1</sup>prot) M2 : 148,7 ± 72,6 vs 65,6 ± 18,9, p < 0.05; Bmax M3: 226,9 ± 77,3 vs 88,6 ± 30,1, p < 0.05); the severity of the phenylephrine induced bradycardia was correlated with the density of the muscarinic receptors. In addition, those rabbits displayed an acetylcholine esterase (AChE) mRNA amplification ratio of 3.6 in heart cells versus normal rabbits. Seeking blood markers, we found that M2 mRNA expression level in lymphocytes was about 10 times higher in our VHR model compared to that observed in normal rabbits, and that it was associated with twice the AChE enzyme activity in erythrocytes.

In a second study, we aimed to seek biological abnormalities in the peripheral vago-cardiac system in SIDS victims. Left ventricular samples were obtained from autopsies of SIDS (n = 9) and children deceased from non cardiac causes (n = 11). Binding experiments performed with a selective muscarinic ligand showed that the muscarinic receptor density in the heart of SIDS victims was more than double that found in samples from children who died from other causes.

**In conclusion:** we showed that overexpression of cardiac muscarinic receptors may play a critical role in a VHR animal model and in infants deceased from SIDS. Furthermore, a same pattern of changes was detected in peripheral mononuclear white blood cells in rabbits. Muscarinic receptor expression level in peripheral mononuclear white blood cells could become a reliable and easily measurable marker of risk of vasovagal syncope and sudden death which could be of great clinical interest.

#### O6-6

##### Exploring a novel molecular mechanism underlying the cardiac development implicated in the outflow tract defects

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The genetic basis of most congenital heart defects (CHD) is still largely unknown. A genetic interaction of two progenitor cell lineages, the cardiac neural crest (CNC) and the second heart field (SHF), play key roles in development of the cardiac outflow tract (OFT). In order to explore molecular mechanisms underlying OFT defects, we analyzed genes essential for CNC and SHF, using DNA from patients with OFT defects, and identified a transcription factor GATA6 as a novel genetic cause of OFT defects. We also demonstrated that Gata6 directly regulated the expression of Semaphorin 3C (*Sema3c*) that mediates a neurovascular guiding signaling essential for the interaction between CNC and SHF, and that mutations in *GATA6* disrupted its direct regulation of *SEMA3C*, resulting in OFT defects. To further clarify the regulatory mechanism of *Sema3c*, we delineated putative promoter/enhancer sequences of *Sema3c* and identified conserved regulatory elements for Fox and Sox factors in the 5'UTR region, and for T-box factors in the 3'UTR region, respectively. Among Fox, Sox and T-box transcription factors, Foxc1, Foxc2, Sox4 and Tbx1 have been shown to play essential roles in the OFT development. Interestingly, Foxc1, Foxc2 and Sox4 showed positive transactivation of *Sema3c* through direct bindings to their regulatory elements. Moreover, Tbx1 negatively regulated the transactivation of *Sema3c* by Gata6, probably via its direct binding to the regulatory elements on the 3'UTR sequence of *Sema3c* along with its direct interaction with Gata6. Consistent with these results, in transgenic mice where the *lacZ* reporter is expressed under control of the 3'UTR enhancer of *Sema3c*, the *lacZ* expression was upregulated and ectopically expanded to whole pharyngeal mesenchyme in mice with *Tbx1* hypomorphic alleles, while it was restricted to the SHF in the pharyngeal mesoderm of wild-type mice. These results suggest a novel molecular mechanism underlying the differentiation of OFT progenitor cells where Tbx1 may restrict the *Sema3c* expression to progenitor cells in the SHF and Gata6, Foxc1/c2 and Sox4 may activate the *Sema3c* expression in these cells in a process of their migration and differentiation into OFT myocardium, eventually leading to a proper signal for the CNC migration into the OFT.

#### O7-1

##### Four-dimensional velocity encoded magnetic resonance imaging improves blood flow quantification in congenital heart disease

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**Purpose:** Valvar stenosis causes accelerated and often complex non-laminar flow patterns that are difficult to assess quantitatively using two-dimensional velocity encoded magnetic resonance imaging (2D VEC MRI). We sought to evaluate the use of 4D VEC MRI for visualizing flow patterns and measuring flow velocities and volumes in complex flow conditions as frequently encountered in congenital heart disease.

**Materials and Methods:** Peak velocities (Vmax) and flow volumes (SV) were quantified by 2D and 4D VEC MRI in volunteers (n = 7) and patients with aortic or pulmonary valve stenosis



( $n = 17$ ). Measurements were performed at standard planes (=level 1) by both techniques and at three further levels in the ascending aorta (levels 2–4) and two in the pulmonary trunk (levels 2–3) using 4D VEC MRI. In patients, regions of peak flow velocities were visualized by 4D VEC MRI and targeted for blood flow quantification. In all patients Vmax was also measured by Doppler echocardiography.

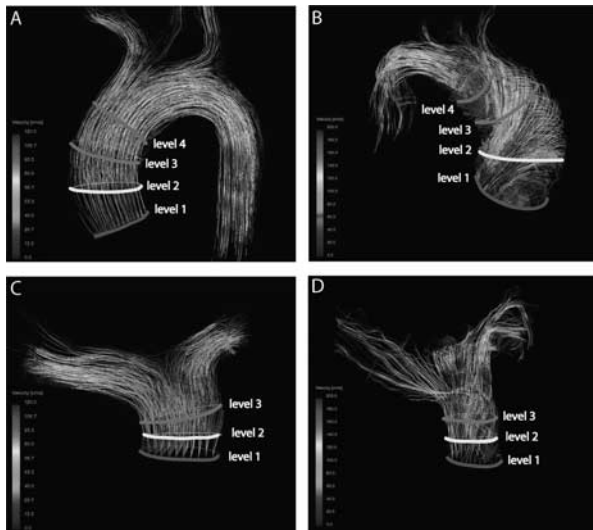


Figure: In healthy volunteers flow patterns along the ascending aorta and main, right and left pulmonary artery were laminar in part swirling but not turbulent (Figure A,C). In patients with aortic or pulmonary valve stenosis flow patterns showed a large variance with vortex formation, helical blood flow and presumably local turbulences (Figure B,D).

**Results:** In volunteers flow patterns were laminar. In patients there were complex flow patterns characterized by high velocity jets and vortex formation. In volunteers and patients, SV at level 1 were comparable between 2D and 4D VEC MRI, but Vmax was higher using 4D VEC MRI ( $p < 0.03$ ). When comparing flow as measured at different anatomical levels in the ascending aorta, there was a significant larger variance in SV in patients than in volunteers ( $p = 0.004$ ). Peak flow velocity visualized by 4D VEC MRI correlated better with Doppler echocardiography than 2D VEC MRI ( $r = 0.7$  vs.  $r = 0.5$ ).

**Conclusion:** 4D VEC MRI, like Doppler echocardiography, can target and thus improve the assessment of peak flow velocity in valvar stenosis. In addition, 4D flow allows visualizing regions with very complex flow that might be associated with turbulences. Therefore, measurement accuracy can be potentially improved.

## O7-2

### Effect of age on exercise capacity and cardiac reserve in patients with pulmonary atresia with intact ventricular septum after biventricular repair

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**Introduction:** In patients with pulmonary atresia with intact ventricular septum (PAIVS) biventricular repair appears to be the optimal treatment option in the absence of significant right

ventricular (RV) hypoplasia. However, long term clinical outcome studies are limited. The purpose of this study was to evaluate exercise capacity and cardiac function during stress in children and young adults with PAIVS.

**Methods:** Ten PAIVS patients after biventricular repair, age  $20.4 \pm 3.7$  years, range nine to 43 years, underwent a cardiopulmonary exercise test, dobutamine stress magnetic resonance imaging (DS-MRI) and delayed contrast enhancement MRI. The response to physical and pharmacological stress and presence of myocardial fibrosis were evaluated and the correlation between patients' age and cardiac response to the physical and pharmacological stress was assessed.

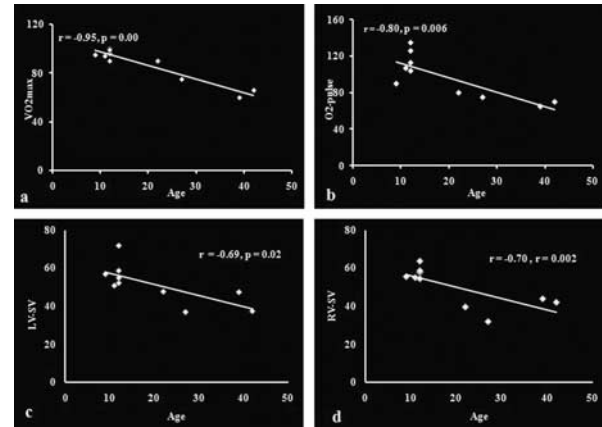


Figure. 1 Correlation between the patients' age and cardiac work indices in response to physical stress (a)  $VO_2\max$ , (b)  $O_2$ -pulse. Correlation between the patients' age and biventricular SV response to pharmacological stress, (c) LV-SV, (d) effective RV-SV.

**Results:** There was a strong negative correlation between the patients' age and physical exercise capacity (Figure 1-a, 1-b). RV E/A volume ratio at rest was negatively correlated with the patients' age indicating impaired RV diastolic function with age ( $r = -0.70$ ,  $p = 0.02$ ). The patients' age was negatively correlated with biventricular stroke volume (SV) response to pharmacological stress (Figure 1-c, 1-d).  $VO_2\max$  and  $O_2$ -pulse during physical stress were strongly correlated with biventricular SV response to pharmacological stress.  $\Delta RV-SV$  correlated with RV E/A volume ratio ( $r = 0.58$ ,  $p = 0.01$ ).

**Conclusion:** In PAIVS patients after biventricular repair exercise capacity and cardiac reserve decrease with age. This findings appears to be related to impaired diastolic RV function and decreased RV filling during stress, indicating that the function of the relatively small RV in PAIVS deteriorates in time.

## O7-3

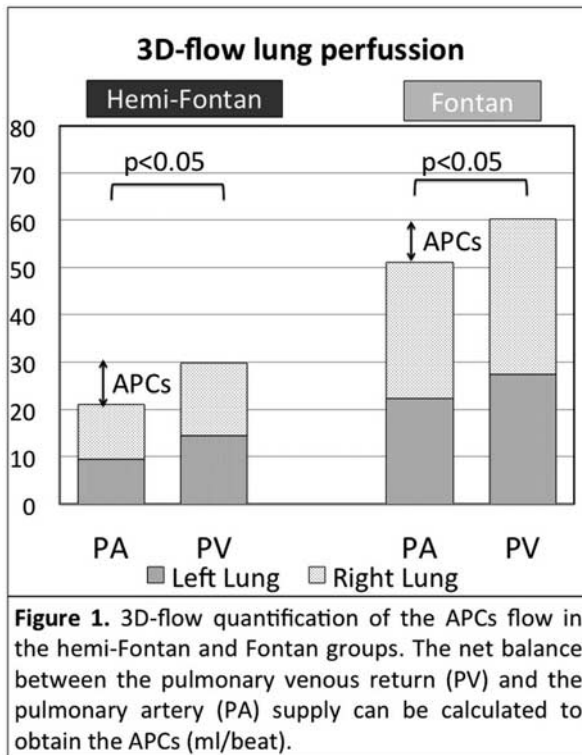
### Quantitative blood flow analysis of aorto-pulmonary collaterals by 3D-flow Phase-Contrast Magnetic Resonance Imaging in patients with single ventricle physiology

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**Introduction:** Aorto-pulmonary collaterals (APCs) have been associated with increased morbidity in patients after single ventricle circulation palliation surgery. Diagnosis by invasive angiography is

qualitative only. Whilst cardiovascular magnetic resonance (CMR) using two-dimensional time-resolved flow (2D-flow) at multiple sites has been shown to successfully quantify APC flow; disadvantages however include complexity requiring very experienced CMR operators, and potential for slice malposition and hence inaccurate flow quantification. CMR three-dimensional time-resolved flow (3D-flow) might be an elegant solution to this problem.



**Materials:** Twenty patients were included in this two-centre prospective study to quantify APCs flow, 10 with previous hemi-Fontan surgery ( $2.5 \pm 1$  years, mean  $\pm$  standard deviation) and 10 with Fontan surgery completion ( $14 \pm 6$  years). Five 2D-flow acquisitions were performed in the aorta, superior and inferior vein cava, right and left pulmonary arteries to serve as gold-standard. They were compared with flow results from a single whole-heart 3D-flow acquisition. In each patient, the 2D flow planes were registered into the 3D flow volume data set to allow accurate alignment of all 5 positions.

**Results:** 3D-flow and 2D-flow results showed good agreement in all investigated vessels (Bland-Altman, mean difference 1.1 ml, limits of agreement  $-7.1$  to  $9.3$  ml). Single 3D-MR-flow was faster and easier compared with the five 2D-flow measurements (12:14 min of 3D-MR-flow vs. 12:58 min for 2D-flow scan-time plus another  $\sim 5$  min of 2D planning,  $p < 0.05$ ).

Table 1. Pulmonary to systemic ratio (Qp:Qs). Q<sub>PA</sub>: Flow pulmonary artery. Q<sub>AO</sub>: Flow aorta. Q<sub>PVR</sub>: Flow pulmonary venous return. Q<sub>SVR</sub>: Flow systemic venous return.

	Qp:Qs without APCs (Q <sub>PA</sub> :Q <sub>AO</sub> )	Qp:Qs with APCs (Q <sub>PVR</sub> :Q <sub>SVR</sub> )	p value
Hemi-Fontan	0.46 : 1	0.80 : 1	0.01
Fontan	0.79 : 1	1.23 : 1	<0.01

On the basis of this validation, to investigate the APCs flow we compared the pulmonary inflow via branch pulmonary artery

supply with pulmonary outflow via all pulmonary veins. It revealed a significant increase in the lung perfusion due to the APCs of  $6 \pm 10$  ml in the hemi-Fontan and  $9 \pm 15$  ml in the Fontan group ( $p < 0.05$ , Figure 1).

The pulmonary to systemic ratio (Qp:Qs) including the APCs (hemi-Fontan 0.8:1, Fontan 1.23:1) was significant different than without including the APCs (hemi-Fontan 0.46:1, Fontan 0.79:1) ( $p < 0.05$ , Table 1).

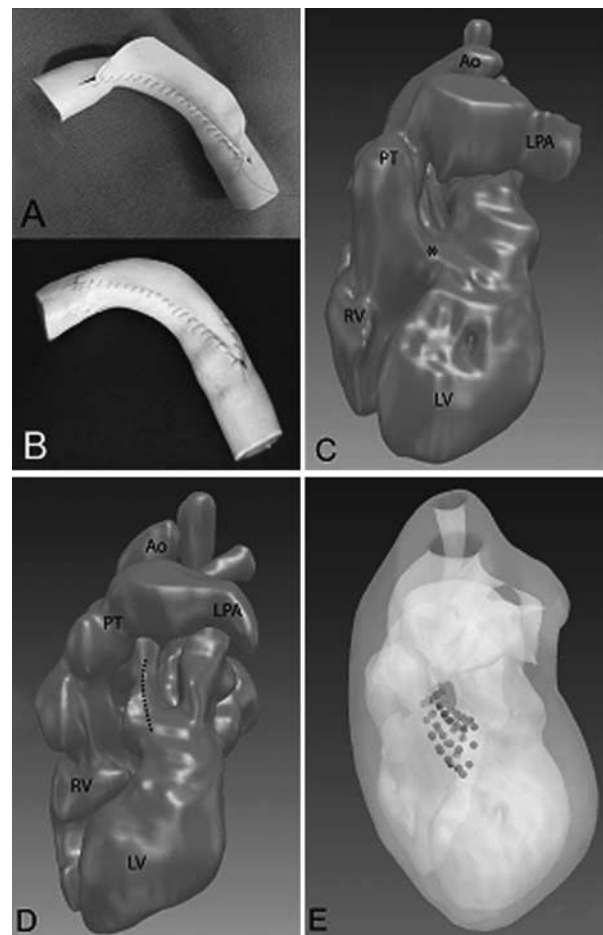
**Conclusion:** Single 3D-flow is accurate compared with multiple-site 2D-MR-flow, but easier to plan, faster to scan and provides further flow information from additional sites. This allows to reliably quantify APC flow in patients with single-ventricle physiology. The method has potential to become clinically valuable in routine clinical follow-up.

#### O7-4

##### Computer aided planning of patches and conduits for surgery in congenital heart disease

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**Introduction:** To optimize outcome in congenital heart disease, the aim is to establish computer assisted methods for planning and simulating surgery in an objective and quantitative manner.



**Methods:** Based on 3D MRI datasets, two applications were developed: (1) for surgical correction of hypoplastic aortic arches the calculation of vessel diameters, and a patch for optimal

surgical correction. This application was evaluated on 12 test datasets and phantoms. (2) For assessing the feasibility of biventricular repair in the case of complex cardiac malformations, an application for simulation of intracardiac repair with a Rastelli like procedure was developed and tested.

**Results:** (1) In all test datasets with varying aortic arch pathologies, diameters were determined with the new application with minor differences ( $1.5 \pm 1.2$  mm) compared to the standard measurements. Individual patches were calculated. In phantoms (Goretex) of pathologic aortas, the patches were inserted successfully (Figure: Goretex phantom with partial (A) and fully (B) inserted patch). MRI thereafter revealed well-formed aortas.

(2) Furthermore, an intracardiac conduit was computed for biventricular repair (Figure, panel C, D and E).

**Conclusions:** The calculation of patch material for surgical reconstruction of aortic pathologies is possible from 3D MRI data and first preclinical tests were successful. For complex cardiac malformations, the preoperative evaluation of the operation method will be feasible. Further preclinical testing is needed.

#### O7-5

##### Speckle tracking in hypoplastic left heart syndrome: correlation with magnetic resonance imaging and the impact of ventricular morphology

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King's College London, UK (1); Evelina Children's Hospital, Guy's and St Thomas' NHS Trust, London, UK (2)

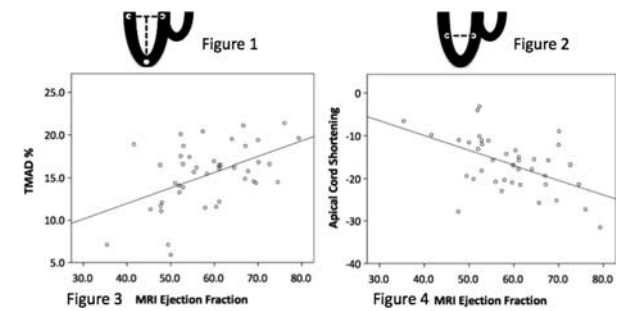
**Introduction:** Assessment of function in the systemic right ventricle (RV) is challenging due to the complex geometry. For the single ventricle heart in Hypoplastic Left Heart Syndrome (HLHS) the current gold standard for assessment of systolic function is cardiac magnetic resonance imaging (MRI) which typically requires general anaesthesia in children under 10 years and is relatively expensive. A simple, quick and reproducible echocardiographic method to assess RV function in HLHS would be ideally suited to the clinical identification of myocardial dysfunction.

**Methods:** Ethical and institutional approval was obtained. Patients with HLHS undergoing cardiac MRI underwent an echocardiogram during the same general anaesthetic. Standard apical four chamber views were acquired using a size appropriate probe (Philips IE33 ultrasound system). Tricuspid annular descent (TAD) was calculated using speckle tracking (figure 1). Two points were placed either side of the tricuspid valve annulus and a third at the apex. TAD was expressed as a percentage of the distance between the tricuspid annular plane and the apex. Speckle tracking derived shortening of a chord placed on the 4 chamber view (figure 2) horizontally in the midcavity region was calculated. Both were then correlated with MRI ejection fraction (EF) (from disc summation from cine images). Left ventricular (LV) morphology and operative Stage were recorded.

**Results:** 43 patients had TAD and horizontal chord shortening. Overall there was a significant correlation ( $p < 0.001$ ) with MRI EF for both (figures 3 and 4). By LV morphologic type, TAD correlation was only significant in the globular LV group. Chord shortening was significant in both the globular and borderline LV groups. TAD correlation was only significant after Norwood Stage 1 whereas chord shortening was significant after both Stage 1 and 2.

**Conclusions:** Significant correlations were apparent in these rapid, simple analyses performed on a standard view. Although both correlated significantly overall, the strength of the correlation was dependent on LV morphology and operative stage. In the few

patients in our cohort with poor function, both TAD and horizontal chord shortening appeared consistently low, suggesting these tools potentially could be used as a screening tool to elicit patients who need further investigation.



#### O7-6

##### Mechanisms of Tricuspid Valve Regurgitation in Hypoplastic Left Heart Syndrome: A Case-Matched Study

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**Background:** Tricuspid valve (TV) insufficiency is associated with poor outcomes in patients with hypoplastic left heart syndrome (HLHS) undergoing staged palliation. Mechanisms of valvular regurgitation in these patients are poorly understood.

**Methods:** We reviewed all patients with HLHS undergoing staged palliation and TV repair from 1998–2008. 2D echocardiograms were retrospectively reviewed by two blinded observers. Structural abnormalities of the TV, including annular dilatation, prolapse, chordal elongation/deficiency, restriction, dysplasia and papillary muscle abnormalities were assessed. Mechanisms of TV insufficiency were categorised. Each patient (“Case”) undergoing TV repair was matched to a control patient, using diagnosis, ventricular function, palliation type, weight and BSA. Variables were analysed in a logistic regression analysis. Level of agreement between observers was evaluated using kappa coefficient.

**Results:** Thirty-four HLHS patients underwent TV repair at median age 6.8 (range 0.3–209) months and median weight 5.8 (range 3.0–50.7) kg. Every valve repair patient had abnormalities of  $\leq$  leaflet versus 14 (44%) controls ( $p < 0.001$ ). The most important cause of TV was leaflet prolapse, followed by leaflet restriction (Table 2). Most commonly affected leaflet was the anterior in 33 patients (97%), the septal was abnormal in 31 (91%) and the posterior leaflet in 23 (68%). Interobserver agreement was excellent ( $\kappa$  values 0.64–0.88). Using the logistic regression analysis, no individual abnormality was predictive of TV repair.

Table 1

	TV repair patients n (%)	Controls n (%)
Leaflet prolapse	23 (67.6)	14 (41.2)
Leaflet restriction	21 (61.8)	11 (32.4)
Leaflet dysplasia	13 (38.2)	0
Valvar/subvalvar thickening	16 (47.1)	6 (17.6)
Chordal elongation	20 (58.8)	8 (23.5)
Chordal deficiency	1 (2.9)	0
Papillary muscle abnormality	3 (8.8)	1 (2.9)
Endocardial fibroelastosis	6 (17.6)	14 (41.2)
Ventricular dilatation	34 (100)	17 (50.0)
Annular dilatation	23 (67.6)	14 (41.2)



Table 2

	Primary Mechanism of Regurgitation n (%)	Secondary Mechanism of Regurgitation n (%)
Leaflet prolapse	20 (58.8)	5 (14.7)
Leaflet restriction	2 (5.8)	13 (38.2)
Annular dilatation	9 (26.5)	5 (14.7)
Leaflet dysplasia	3 (8.8)	5 (14.7)

**Conclusions:** Important structural abnormalities of the TV are common in patients with HLHS and TV insufficiency, and may be readily identified using 2D echocardiography.

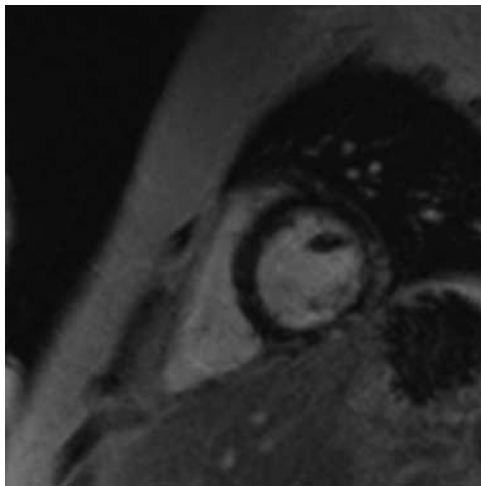
### O7-7

#### Cardiac involvement in Duchenne Muscular Dystrophy (DMD) – a detailed cardiac magnetic resonance imaging study in 39 patients

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**Background:** In the second decade of life about 70% of DMD patients develop initially asymptomatic cardiomyopathy with impairment of left ventricular function, which is one of the major reasons for subsequent morbidity and mortality in DMD. The value of cardiac resonance imaging (CMR) regarding sensitivity and significance in the diagnosis of cardiac involvement is to be evaluated.

**Methods:** CMR including late gadolinium enhancement (LGE) sequences as well as standard cardiologic diagnostics and Tissue Doppler Imaging were performed in 39 male DMD patients (mean age 13 years, range 6–20 years). We here focus on MRI analysis.



**Results:** 16 patients (42%) had a limited left ventricular ejection fraction. Left ventricular mass was reduced without dilation; diminished end diastolic volume was present in 14 patients (37%), only one patient had a dilated left ventricle. CMR showed regional wall motion abnormalities in 47% of the patients. LGE was detectable in 89%, preferentially in the inferolateral and anterolateral segments of the left ventricle. The average mass of LGE was 7 g/m<sup>2</sup> (range 0–29 g/m<sup>2</sup>) with a mean fraction of 14% of left ventricular mass (0–51%). The youngest patient with evidence of LGE was five years old. LGE correlated with the wall motion abnormalities in CMR and Tissue Doppler Imaging.

**Conclusions:** In our group of 39 Duchenne patients, cardiomyopathy manifested with a left ventricular muscular atrophy without dilation. Localised LGE marks the extent of inflammation and fibrotic tissue transformation. Regional impairment of wall motion starts at an early stage. Further and ongoing investigations will show, if regional myocardial impairment and LGE in CMR analysis are reliable indicators for early medical treatment.

### O8-1

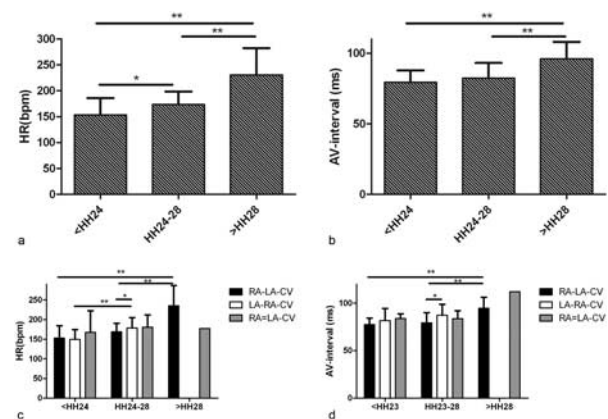
#### Development of the sinoatrial and atrioventricular nodes in the avian embryo: a reference series of morphological and electrophysiological changes during maturation

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**Objectives:** Analysis of the morphological and electrophysiological development of the sinoatrial (SAN) and atrioventricular (AVN) nodal areas can help us to unravel the mechanisms behind specific arrhythmias. In this study we provide an overview of the morphological changes that occur in SAN and AVN differentiation during avian heart development, and provide baseline measurements of developmental changes in the electrograms.

**Methods:** The developing SAN and AVN area were studied for expression patterns of the cardiac markers cTnI, Nkx2.5, the gap junction protein Cx43 and the cation channel HCN4. The developing electrogram was studied by *ex ovo* local electrophysiological recordings as well as atrial activation patterns.



**Figure.** Electrophysiological changes during development. a-b show the changes in heart rate (HR) in beats per minute (bpm) (a) and atrioventricular (AV) interval in milliseconds (ms) (b) during development. c-d show the changes in HR and AV interval during development for the different atrial activation patterns. RA-LA-CV: right-sided dominant pacemaker; LA-RA-CV: left-sided dominant pacemaker; RA=LA-CV: concurrent activation. \*:p < 0.05; \*\*:p < 0.005.

**Results:** Initially, the entire sinus venosus myocardium expresses cTnI, HCN4, but not Nkx2.5, and has the potential to generate the first electrical activity resembling a pacemaker. At later stages both expression patterns and electrical activation patterns become restricted to the definitive right sided SAN. Similarly,

the early atrioventricular canal and at later stages the atrioventricular ring myocardium and AVN areas show a common expression pattern. During development we observed a significant increase in heart rate and atrioventricular delay (Figure). Lineage tracing experiments show a potential sinoatrial contribution to the AVN area.

**Conclusions:** Significant changes occur in both morphology and electrical properties of chick sinus venosus and atrioventricular ring myocardium during development, where the putative SAN and AVN will form, respectively. The broad electrical potential of these structures during development may form an explanation for the occurrence of predilection sites for arrhythmias in the adult.

## O8-2

### Pathologic anatomy of the coronary sinus and cardiac veins in double discordance (congenitally corrected transposition of the great arteries)

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Double discordance (DD) is a rare congenital heart defect (CHD) associating discordant atrioventricular and ventriculoarterial connections. Late prognosis in physiologically corrected or unoperated DD depends on the progressive failure of the systemic right ventricle (RV). A possible cause for systemic RV dysfunction could be ventricular dyssynchrony. Cardiac resynchronization therapy (CRT) may thus be indicated in some cases. However, the uncertainties about anatomy of the coronary sinus (CS) and cardiac veins often prevent the cardiologists from implanting a 3-lead CRT transvenous system in these patients.

**Aim of the study:** To evaluate the anatomy of the CS and cardiac veins in specimens with DD, in order to assess the feasibility of transvenous CRT.

**Material and Methods:** Among the 1337 heart specimens available in the anatomic collection of the French Center of Reference for complex CHD, 22 had DD with 2 ventricles. Hearts were reviewed with special attention paid to the course and drainage of the CS and cardiac veins. Segmental anatomy, location of the VSD, status of the pulmonary outflow tract and anomalies of the atrioventricular valves were reviewed.

**Results:** Segmental anatomy was S,L,L in 20/22 hearts, S,L,D and I,D,D in 2. There was a VSD in 21/22, pulmonary atresia in 9, subpulmonary stenosis in 3, abnormal tricuspid valve in 17/22 including Ebstein anomaly in 6, straddling in 7. The CS was always located behind the morphologically left atrium (LA). However, its anatomy was normal, with normal drainage into the morphologically right atrium, in only 13/22 (59% of cases). The CS was of reduced length with normal orifice in 6. Orifice was atretic in 3 (1 completely absent CS with direct drainage of coronary veins into the LA, 2 with normal size CS). At least 1 available vein was found in all cases with patent CS orifice.

**Conclusion:** CS in DD is always located behind the morphologically LA. However, its anatomy is abnormal in 41% of cases. The most frequent anomalies are reduced length (27%) and atretic orifice (14%). This advocates the use of imaging techniques (multislice CT imaging or CS venography) before considering transvenous CRT in these patients.

## O8-3

### Permanent Cardiac Pacing in Children – Choosing the Optimal Pacing Site: A Multi-Center Study

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**Objectives:** We sought to evaluate the effects of ventricular pacing site on LV synchrony and function in children requiring permanent pacing.

**Methods:** 152 children and adolescents (17 centers) with complete AV block and a structurally normal heart undergoing permanent pacing were prospectively studied. Median age was 11.2 yrs (interquartile range (IQR) 6.6–15.3). Median pacing duration was 5.3 yrs (IQR 3.0–8.6). Data were analyzed in a core lab. Pacing sites were the free wall of the RV outflow tract (RVOT, N = 11), lateral RV (RVLat, N = 31), RV apex (RVA, N = 58), RV septum (RVS, N = 18), LV apex (LVA, N = 11), LV mid-lateral wall (LVLat, N = 16) and LV base (LVB, N = 7).

	RVOT	RVLat	RVA	RVS
IVMD ms	35 (17/59)	55 (34/63)	41 (23/59)	42 (32/48)
SPWMD ms	83 (70/90)	70 (50/80)	40 (0/70)	90 (78/90)
LVDI ms	189	163	87	134
LVEF %	47 (46/53)	52 (43/56)	54 (51/59)	52 (46/57)
	LVA	LVLat	LVB	P<
IVMD ms	9 (-2/17)	-12 (-27/-3)	-16 (-31/-4)	0.001
SPWMD ms	0 (-30/0)	-30 (-75/0)	-90 (-90/60)	0.001
LVDI ms	44	73	136	NA
LVEF %	58 (55/61)	62 (57/66)	55 (51/64)	0.001

**Results:** Pacing sites differed in inter-ventricular mechanical delay (IVMD), septal to posterior wall motion delay (SPWMD), LV dyssynchrony index (sum of absolute mechanical delays from LV radial strain, LVDI) and LV ejection fraction (EF) with LVA/LVLat being superior (Table, median and IQR). LVDI correlated negatively with LVEF (R = 0.85, P = 0.016). Pacing from RVOT/RVLat (OR 7.9, CI 2.1–29.6, P = 0.002) was a significant multivariable predictor of LVEF < 45%. LVA/LVLat pacing (OR 8.7, CI 2.5–30.5, P < 0.001) predicted preserved LV function

(LVEF  $\geq$  55%). Presence of maternal anti Ro/La antibodies, age at implantation, pre-implantation LV size and function, duration of pacing, DDD mode and QRS duration had no significant impact. **Conclusions:** LV mechanical synchrony and function may significantly deteriorate with RVOT/RVLat and is best preserved by LVA/LVLat pacing. Although inferior to LVA/LVLat, RVA pacing is well tolerated in the majority. RVS did not show any advantage over RVA pacing. (JJ supported by the research project of Univ. Hosp. Motol MZOFNM2005).

#### O8-4

##### **Cardiac Resynchronization Therapy in Pediatric and Congenital Heart Disease Patients: A Long-Term Single Center Experience**

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**Introduction:** Cardiac resynchronization therapy (CRT) is a promising treatment for selected young patients with systemic ventricular dysfunction. The long-term benefits of this therapy are as yet unknown.

**Methods:** A retrospective single center review of our experience with pediatric CRT.

**Results:** A total of 25 patients underwent CRT since 2003, as adjunct to optimal heart failure therapy. Six patients had CRT as preemptive therapy and were excluded from data analysis. The median age at time of CRT was 9.2 years (range 1 to 24 years). Mean (SD) duration of follow-up was 3.4 (2.2) years. The diagnosis was congenital heart disease (CHD) in 10 (52.6%) patients including 3 patients with systemic right ventricle and 1 with single ventricle. Cardiomyopathy was present in 8 patients (42.1%) and congenital complete heart block in 1 patient (5.3%). 13 patients (68%) had permanent pacemaker implanted prior to CRT and 4 had a combined CRT and implantable cardioverter-defibrillator. Three patients had concurrent cardiac surgery with CRT. Patients underwent dyssynchrony assessment by tissue Doppler for optimization of CRT. Among 19 patients with established ventricular dysfunction at the time CRT was initiated, the mean (SD) baseline (pre-CRT) QRS duration was 139.7 ms(30.1). The mean (SD) pre-CRT systemic ventricular ejection fraction (EF) was 30.7% (11.4), 37.5% (10.5) after a mean follow-up duration of 0.5 (0.4) years and 48.2% (10.7) after a mean of 3.4 (2) years of follow-up ( $p < 0.006$  by paired comparison with baseline value). The baseline and latest EF did not differ according to the underlying diagnosis. Two patients died at 6 weeks and 4 months post CRT and no patient has so far undergone heart transplantation. Three (15.8%) patients were non-responders based on lack of improvement in EF. Five patients needed revision of their CRT device. Survival after CRT was 88.8% at 1 year, 85.7% at 2 years and 83.3% at 4 years.

**Conclusions:** CRT in selected children with systemic ventricular dysfunction can be associated with long-lasting benefit, including survival and improved ventricular function. The benefits are apparent in children with structurally normal hearts and those with congenital heart disease.

#### O8-5

##### **Acute and Long-Term Outcome after Catheter Ablation of Supraventricular Tachycardia in Patients after the Mustard or Senning Operation for D-Transposition of the Great Arteries**

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**Background:** Data about the long-term outcome of catheter ablation in patients (pts) with D-Transposition of the great arteries (d-TGA) after the Mustard/Senning operation are scarce. **Methods:** We retrospectively evaluated the 27 pts (mean age  $28.7 \pm 6.7$  years, 9 females) after a Mustard ( $n = 16$ ) or Senning ( $n = 11$ ) procedure who underwent ablation for supraventricular tachycardia at our institution from January 2004 to July 2010. A 3D mapping system (Carto) was used in all cases (in combination with remote magnetic navigation (RMN) in 14 cases). Tachycardia mechanism was atrial tachycardia (AT) including intraatrial reentrant tachycardia/focal atrial tachycardia or atrioventricular node reentrant tachycardia (AVNRT). Acute ablation success was defined as termination of AT or non-inductivity of AVNRT after ablation. Follow-up was available 3, 6 and 12 months after ablation and yearly thereafter. Long-term success was defined as freedom from tachycardia.

**Results:** In the 27 pts (AT  $n = 22$ , AVNRT  $n = 2$ , AT+AVNRT  $n = 3$ ), 35 procedures were performed (one procedure  $n = 20$ , two  $n = 6$ , three  $n = 1$ ). Overall 36 tachycardia forms were found. Tachycardia mechanism included AT ( $n = 31$ ) and AVNRT ( $n = 5$ ). Tachycardia was ablated manually ( $n = 27$ ) or using RMN ( $n = 9$ ) with an acute success in 31/36 (86%) tachycardias. After the first ablation, recurrence occurred (AT  $n = 8$ ; AVNRT  $n = 0$  pts). After a mean of 1.3 ablations and a mean follow-up time of  $29.1 \pm 24.5$  months, 26/27 (96.2%) patients were free from AT/AVNRT. Acute and long-term success in the last 9 procedures (during the years 2008–2010) using exclusively RMN was 100%.

**Conclusions:** Catheter ablation of AT or AVNRT in patients after the Mustard or Senning operation for d-TGA has a high acute success rate and with the use of RMN long-term results are excellent after a single ablation.

#### O8-6

##### **Genetic background of long QT syndrome in infants, children, and adolescents in Japan**

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**Introduction:** Genetic studies for long QT syndrome (LQTS) have reported that prevalence of LQT1, LQT2, and LQT3 among these three mutations in adults is approximately 50%, 40%, and 10%, respectively. However, little data are available for pediatric population. In 1994, a school-based ECG screening program was started for all 1st, 7th, and 10th graders in Japan. The program screened children with definite LQTS without symptoms. We aimed to determine the characteristics of genetic background in childhood LQTS in Japan.

**Methods:** The study population included 170 family members from 81 probands (0–18 years; median, 9.5 years; M:F = 45:36) who were referred to our center from 1993–2010. Probands had a QTc value of  $0.45^{0.5}$ . Genomic DNA was isolated from blood and screening for LQT1–LQT8, except for LQT4, was re-performed by PCR and direct DNA sequencing. When multiple mutations were present, each mutation was counted in the number of each genotype.

**Results:** Genotypes were identified in 49 of 81 probands (60%) and in 84 of 170 family members. LQT1 was found in 43 family members (from 24 probands), LQT2 in 21 (15), LQT3 in 17 (9), LQT5 in seven (3), LQT7 in four (2), and LQT8 in one (1). Double mutations or gene variants were found in eight subjects and triple mutations or gene variants were found in one subject. The prevalence of LQT3 among the three main genotypes in probands (9/44, 20%) and in family members (17/77, 22%) in



the present study was significantly higher (both  $p = 0.004$ ) than the data for the adult population (11/192 in probands and 82/812 in family members by Sauer AJ, et al. in 2007). Of 81 probands, the screened subjects showed a higher rate of genotypic determination (27/33) than symptomatic subjects (17/37,  $p = 0.003$ ) or a miscellaneous group including family examinations (5/11,  $p = 0.045$ ). The QTc values were not different between the screened and symptomatic groups.

**Conclusions:** A high prevalence of the LQT3 genotype in the pediatric population suggests progress in the medical management of these patients during infancy and childhood. School-based ECG screening and genetic testing may help prevention of LQTS-related symptoms in Japan.

#### O8-7

##### **Predictors of Adverse Events in Children with Arrhythmogenic Right Ventricular Cardiomyopathy**

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**Introduction/Objective:** Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an increasingly recognized cause of sudden death in young adults and children. Children may be referred for symptoms, signs or a positive family history of ARVC or an ARVC-causing gene. We sought to identify predictors of adverse events among referred children meeting 2010 proposed modified Task Force criteria for probable or definite ARVC.

**Methods:** We reviewed all patients referred to a pediatric cardiovascular genetics/electrophysiology program for assessment of possible ARVC. Patients were routinely investigated with ECG, SAECG, Holter, Echo MRI and (after 2003) gene testing. If only partial criteria for ARVC were present, patients underwent catheterization for RV angio, EPS and biopsy. Adverse events were defined as sustained VT, appropriate ICD discharge or death. Univariate predictors of adverse events were assessed by unpaired T-test, Chi-square analysis or Fisher's Exact Test as appropriate.

**Results:** Of 423 patients referred for assessment, 77 met inclusion criteria (48 ARVC, 29 probable ARVC) with a mean age of  $14.9 \pm 3.3$  (range 5 to 18) years at last follow-up. No adverse events were identified in 71, whereas 6 had adverse events (including 1 death, 3 appropriate ICD discharges and 3 sustained VT). Predictors of adverse events included presentation as the index case v. relative (6/35 v. 0/35,  $p = 0.026$ ), extra-systoles, (4/21 v. 1/45,  $p = 0.049$ ) prolonged S duration in V1 (73 v. 53 ms,  $p = 0.006$ ), major criteria for global or regional dysfunction (6/32 v. 0/33,  $p = 0.027$ ), documented LBBB VT (4/13 v. 2/57,  $p = 0.020$ ), any minor arrhythmia criteria (5/24 v. 1/44,  $p = 0.031$ ) and absence of any major family history criteria (6/34 v. 0/36,  $p = 0.018$ ). There was a tendency for patients meeting ARVC criteria to have adverse events compared to those diagnosed with borderline criteria.

**Conclusions:** Adverse events can occasionally occur in children identified with probable or definite ARVC. Children presenting with symptoms, signs of right ventricular dysfunction or documented arrhythmias appear to be at highest risk of adverse events, compared to those referred with or identified to have a family history of ARVC. These findings should be validated in a larger cohort or multicenter study of children with ARVC.

#### O9-1

##### **Clinical Characteristics of Pediatric Pulmonary Hypertension: Results from the global registry Tracking Outcomes and Practice in Pediatric Pulmonary Hypertension (TOPP)**

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**Introduction:** Pediatric and adult pulmonary hypertension (PH) share similar pathology and clinical characteristics yet several features appear different. However, pediatric data are lacking. TOPP, a global, observational study, provides demographic, clinical and treatment patterns in pediatric PH.

**Methods:** Consecutive patients are screened and consenting patients with PH (mean pulmonary artery pressure [mPAP]  $\geq 25$  mmHg, pulmonary capillary wedge pressure  $\leq 12$  mmHg and pulmonary vascular resistance index [[PVRi]  $> 3$  WU.m<sup>2</sup>) are enrolled at 31 centers in 20 countries.

**Results:** Of 456 patients (age at diagnosis  $< 18$  yrs), enrolled between January 2008 and February 2010, 362 had confirmed PH. Of the PH-confirmed patients, median age at diagnosis was 7 yrs with a female preponderance (1.4:1). Mean time from symptom onset to diagnosis (17 mos; 95%CI 14–20 mos) was shorter than reported in adults. The majority (88%) had Group 1 PH: pulmonary arterial hypertension (PAH), of which 57% had idiopathic or familial PAH (IPAH/FPAH) and 43% had PAH associated with other conditions (APAH). Of the APAH patients, 85% had associated congenital heart disease (CHD). The remaining children (12%) had PH associated with lung diseases and/or hypoxemia, with bronchopulmonary dysplasia (BPD) the most frequent association. Co-morbid conditions, including chromosomal abnormalities, other syndromes and anomalies, were reported in 24%. As in adults, dyspnea and fatigue were the most frequent presenting symptoms. In contrast to adult PH, syncope was reported frequently, especially in children with IPAH/FPAH (31%) or repaired CHD (18%); in contrast, no children with unrepaired/residual shunts had syncope reported. Functional class (FC) at diagnosis was predominantly FC II (12%/51%/30%/7%, I/II/III/IV, respectively) consistent with preserved right heart function despite severe PAH, i.e. mPAP ( $58 \pm 19$  mmHg) and PVRi ( $16 \pm 10$  WU.m<sup>2</sup>).

**Conclusions:** TOPP demonstrates that pediatric PH has important characteristics that differ from adults, including syncope as a frequent presenting symptom in IPAH/FPAH despite preserved cardiac output. Additionally, the distribution of etiologies, the occurrence of specific associated pediatric conditions, including BPD and a high rate of co-morbidities appear different than in adults. As we track outcomes in these children, we hope to define prognostic parameters for risk stratification to guide therapeutic approaches in pediatric PH.

#### O9-2

##### **Exercise capacity in children with isolated congenital complete atrioventricular block: does pacing make a difference?**

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**Introduction:** The management of patients with isolated congenital complete atrioventricular block (CCAVB) has changed during the last decennia. In the past a minority of patients was paced. The current policy is to pace the majority of patients based on a variety of criteria among which a limited exercise capacity.

Data regarding exercise capacity in this population stems from old publications reporting small case series of unpaced patients. Exercise capacity is an important factor in performing age-appropriate physical activities and future health. Therefore we investigated the exercise capacity of a group of contemporary children with CCAVB.

**Methods:** Sixteen children (mean age  $11.5 \pm 4$ ; 7 boys, 9 girls) with CCAVB were tested. In 13 patients a median number of 3 pacemakers were implanted, while in 3 patients no pacemaker was given. All patients had an echocardiogram and completed a cardiopulmonary cycle exercise test (CPET). Exercise parameters were determined and compared with reference values of healthy Dutch peers.

**Results:**

Variable	CCAVB Group (n = 16)	Paced Group (n = 13)	Unpaced Group (n = 3)
Left ventricular end diastolic diameter	$110 \pm 10$	$108 \pm 10$	$117 \pm 11$
% predicted			
Left ventricular dysfunction	2 patients (13%)	2 patients	0 patient
Interventricular dyssynchrony	8 patients (50%)	8 patients	0 patient
Intraventricular dyssynchrony	1 patient (6%)	1 patient	0 patient
Rest heart rate (HR) in bpm	$73 \pm 13$	$77 \pm 10$	$53 \pm 2$
Peak HR in bpm (% predicted)	$135 \pm 37$ (70 ± 19)*	$139 \pm 35$ (72 ± 18)	$117 \pm 48$ (60 ± 25)
Upper rate behavior (paced patients)	4 patients (25%)	4 patients	n.a.
Peak RER	$1,13 \pm 0,11$	$1,15 \pm 0,12$	$1,06 \pm 0,01$
Peak work load/kg in W/kg	$2,8 \pm 0,6$	$2,8 \pm 0,6$	$3,3 \pm 0,2$
(% predicted)	(91 ± 24)	(87 ± 23)	(112 ± 11)
VO <sub>2peak</sub> /kg in ml/kg/min	$34,4 \pm 9,5$	$33,0 \pm 9,5$	$40,5 \pm 8,3$
(% predicted)	(79 ± 24)*	(75 ± 24)	(93 ± 22)
VAT % predicted	$78 \pm 21*$	$79 \pm 22$	$73 \pm 15$
Peak O <sub>2</sub> pulse in ml/beat	$10,4 \pm 5,1$	$10,8 \pm 5,6$	$8,6 \pm 1,7$
(% predicted)	(111 ± 56)	(98 ± 31)	(170 ± 106)
VE/VCO <sub>2</sub> slope (% predicted)	$33,3 \pm 6,6$ (108 ± 17)	$31,8 \pm 6,3$ (105 ± 18)	$40,0 \pm 1,8$ (121 ± 2)
OUES slope (% predicted)	$1424 \pm 510$ (92 ± 39)	$1513 \pm 528$ (81 ± 23)	$1034 \pm 57$ (141 ± 60)

\*significantly different from normal ( $p < 0,05$ )

**Conclusions:** Children with CCAVB have a significantly reduced exercise capacity compared to healthy peers. Interestingly paced patients did not perform better than unpaced patients. A significant percentage of paced patients showed upper rate behavior of the pacemaker. These results show that CPET can be a valuable tool in detecting suboptimal pacemaker programming.

### O9-3

#### The efficacy and safety of new strategy for refractory Kawasaki disease

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**Background:** Approximately 15–20% of patients with Kawasaki disease (KD) are not responsive to intravenous immunoglobulin (IVIG). This KD was defined as refractory KD. In these refractory KD patients, coronary artery lesions frequently developed. We examined clinical utility and safety of new strategy for refractory KD as IVIG-plus-methylprednisolone combined therapy (IVIG+IVMP) for initial treatment in patients predicted as refractory KD.

**Methods:** Since 2008, 120 KD patients, mean age  $30.5 \pm 22.4$  month were studied at Kitasato University. We predicted refractory KD patients before initial treatment using Egami score (J Pediatr 2006) and randomly divided IVIG + IVMP therapy or single-IVIG treatment to patients. The Egami scoring system identifies age, days of illness, platelet count, C-reactive protein (CRP), alanine aminotransferase (ALT) to predict refractory KD patients before treatment (cut-off: 3 points; 78% sensitivity and 76% specificity).

**Results:** The 45 of 120 KD patients (37.5%) were predicted refractory KD patients using Egami score. The predicted IVIG-responders (n = 75) were administered single-IVIG (2.0 g/kg for 1 day). The 45 patients with prediction of refractory KD were randomly assigned to the single-IVIG group (n = 27) and IVIG+IVMP therapy group (n = 18). The 16 of 18 patients (88.9%) had a prompt defervescence in the IVIG + IVMP therapy group compared with 6 of 27 patients (33.3%) in the single-IVIG group ( $p < 0.05$ ). The 3 patients had coronary artery lesion in the IVIG + IVMP therapy group. However 10 patients had coronary artery lesion in single-IVIG treatment groups. There no serious adverse events in both treatment groups.

**Conclusion:** Our study demonstrated the new strategy for refractory KD as IVIG + IVMP therapy was the effectiveness and safety as a primary treatment for predicted refractory KD patients.

### O9-4

#### The Outcomes of the Extracardiac Conduit Fontan Operation in Patients with Heterotaxy Syndrome: Experience in a Single Institute

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**Introduction:** In patients with single ventricle physiology and heterotaxy syndrome, interim mortality and morbidity before completion of Fontan procedure have been known to be high. We sought to assess the outcomes after completion of Fontan procedure in heterotaxy patients compared with non-heterotaxy patients.

**Methods:** From December 1993 to July 2010, 235 patients have completed extracardiac conduit (ECC) Fontan palliation. We reviewed the results of these patients retrospectively and the data of subgroup having heterotaxy, 57 patients were compared with those who did not have heterotaxy syndrome.

**Results:** 57 patients were comprised of 6 left isomerism and 51 right isomerism. Patients with heterotaxy syndrome had significantly higher number of total anomalous pulmonary venous return (59.6% versus 1.1%) and separate hepatic vein structure (29.8% versus 0.0%). Pulmonary vascular compliance (PA index/mean PAP) were higher in heterotaxy subgroup before Fontan operation. After ECC Fontan procedure, duration of pleural effusion, ICU stay and ventilator use were not significantly different from non-heterotaxy group. In long-term follow-up, grade of NYHA classification, degree of atrioventricular regurgitation, incidence of protein losing enteropathy and reintervention rate were not different in both groups.

Overall mortality rate were 10.1% during 10-year follow up period and there was no significant difference statistically between heterotaxy group and non-heterotaxy group in mortality. (5.1% versus 10.5%,  $p = 0.125$ )

**Conclusion:** Heterotaxy syndrome was not increasing risk of mortality or morbidity once after completion of ECC Fontan operation. This study implies that heterotaxy syndrome has more anomalies in venous connection, however, its complications and prognosis were not different from other functional univentricular heart after Fontan palliation.

#### O9-5

##### **Mild residual pulmonary stenosis after correction of Tetralogy of Fallot is associated with a reduced risk of pulmonary valve replacement during follow-up**

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**Introduction:** Pulmonary valve replacement (PVR) is often performed in corrected Tetralogy of Fallot (ToF) patients with pulmonary regurgitation, in order to reverse right ventricular dilatation and its adverse clinical outcome. Comprehensive knowledge about the predictive value of pre-operative, peri-operative and early post-operative parameters for the risk of PVR during follow-up can be of great value for counselling of patients with ToF and their parents. The aim of this study was to determine the influence of several clinical, surgical and echocardiographic parameters on the need for PVR during follow-up in corrected ToF patients. Specifically, the influence of a residual pulmonary stenosis on the need for PVR was investigated.

**Methods:** The study was a retrospective cohort study. Patients were included if their initial diagnosis was ToF, and if clinical follow-up had taken place at our center for a minimum of five years. Patient characteristics, surgical parameters and post-operative parameters were reviewed, with special focus on the first available data about the presence and severity of post-operative pulmonary regurgitation and stenosis. Univariate and multivariate Cox proportional hazards regression analyses were performed to identify predictors for PVR.

**Results:** A total of 171 ToF patients were included. A total of 71 (41.5%) patients underwent PVR, with a median interval between corrective surgery and PVR of 24.2 years. Multivariate analysis showed that older age at corrective surgery (Hazard Ratio (HR) = 1.20,  $p < 0.001$ ) and the use of a patch (transannular patch HR = 7.55,  $p < 0.001$ , RVOT patch HR = 5.95,  $p < 0.001$ ) significantly predicted PVR during follow-up. Moreover, a mild residual pulmonary stenosis (peak systolic gradient 15 to 30 mmHg) was independently associated with a reduced risk of PVR, as compared with the patients with a gradient  $< 15$  mmHg (HR = 0.47,  $p = 0.020$ ) and those with a gradient  $> 30$  mmHg (HR = 0.37,  $p = 0.022$ ).

**Conclusions:** Besides known risks factors for PVR, such as older age at repair and the use of a patch, post-operative mild residual pulmonary stenosis with a peak systolic gradient between 15 and 30 mmHg is independently associated with a reduced risk of PVR during follow-up of corrected ToF patients.

#### O9-6

##### **Outcome of Acute Myocarditis: A Single-Center 8-year Experience**

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**Introduction:** Acute myocarditis can result in a rapidly progressive and often fatal course in children. There are still very few report regarding the prognosis of this disease. We report our single institutional experience including treatment with extracorporeal membranous oxygenation (ECMO) in children with acute myocarditis.

**Methods:** All patients admitted to our institution with a diagnosis of acute myocarditis from 2002 to 2010 were retrospectively reviewed. The diagnosis of acute myocarditis was made by clinical history, clinical findings, and/or endomyocardial biopsy findings.

**Results:** A total of 30 events in 29 children (one patient suffered twice) were identified with acute myocarditis. The median age was 4 years (range 0 days–11 years). Presenting symptoms included: abdominal pain and/or vomiting (12/30), episodes of change in consciousness (seizures, drowsiness or syncope) (12/30), fatigue (4/30), and upper respiratory symptoms (3/30). The mean left ventricular ejection fraction (LVEF) was 30% at presentation. Intravenous inotropic support was required in all patients and 28/30 were mechanically ventilated. ECMO was implanted in 11 cases. Median duration of device therapy was 6 days (range 3–9 days). The survival rate in patients who required ECMO and in those who did not was 72% (8/11) and 89% (17/19) respectively, resulting in an overall survival rate of 82% (25/30). ECMO was withdrawn in 3/11 with severe brain damage as the indication in all 3 cases. These 3 death cases in the ECMO group all required cardiopulmonary resuscitation prior to ECMO implantation. The remaining 8 cases were all successfully weaned off from ECMO without any major complications. All survivors are currently alive with normal left ventricular function and no neurological sequelae.

**Conclusions:** Despite severe presentation, the outcome of acute myocarditis in children is favourable, with a survival rate of 82% in this series. ECMO is a highly effective form of mechanical circulatory support, and a bridge to recovery can be anticipated in the majority of these patients with excellent prospects for eventual recovery of myocardial function. Early diagnosis and short duration from cardiogenic shock to implantation of ECMO is crucial for further improvement in prognosis.

#### O9-7

##### **Echocardiographic Findings in 30 Children with Various Lysosomal Storage Disease**

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**Background:** Lysosomal storage diseases (LSD) can cause cardiovascular involvement, especially thickening of cardiac valves, ventricular hypertrophy and pulmonary hypertension. Little is known about differences of cardiac involvement in children between various LSD including Fabry disease (FD), mucopolysaccharidosis (MPS), Pompe disease (PD), I-cell disease (ICD) and mucopolipidosis (ML).

**Methods:** The echocardiographic database was searched for all children with the diagnosis of LSD; all echocardiographic data were analyzed as well as clinical findings and information on enzyme replacement therapy (ERT) and bone marrow transplantation (BMT).

**Results:** LSD in children is rare with only 30 patients (19 females) in our echocardiography data base. There were 21 pt with MPS (6 pt with type 1 MPS; 4 pt with type 2 MPS, 3 with type III MPS, 5 pt with type IV MPS, and 3 pt with Type VI MPS), 6 pt with FD, 2 pt with mucopolipidosis III, 1 pt with ICD, and 2 pt



with PD. ERT was given in 5 pt with MPS and 3 pt with FD. Three pt with MPS had BMT.

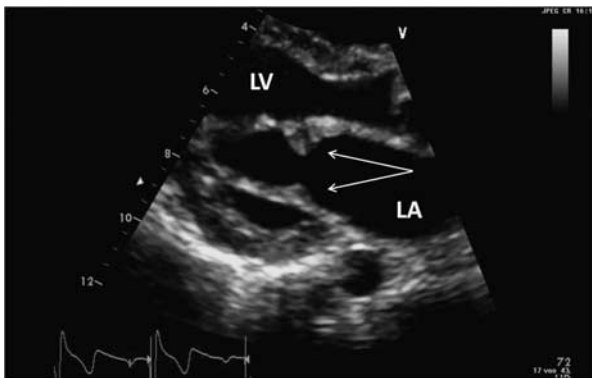
The results of the echocardiographic examinations are shown in the Table. Any valvular heart disease was present in 18 pt. Mitral valve thickening was present in 18 pt, aortic valve thickening in 16 pt. In children valvular heart disease is significantly less often observed in FD than in MPS. Stenosis of the aortic valve was observed in 3 pt, and of the mitral valve in 2 pt (all with MPS). Cardiac symptoms were reported only in 2 pt with MPS. Valvular abnormalities were more common than left ventricular hypertrophy (2 pt).

	No of pt	Age, years	EF, %	LVMMI, g/m <sup>2</sup>
MPS	21	11.2 ± 5.2*	59 ± 4	65 ± 39
Fabry disease	6	8.4 ± 4.3	60 ± 4	67 ± 18
Other	5	4.9 ± 5.3	59 ± 3	48 ± 31*
	LVH	Abnormal mitral valve	Abnormal aortic valve	
MPS	1 (5%)	15 (71%)	14 (67%)	
Fabry disease	0	0	0	
Other	1(20%)	3 (60%)	2 (40%)	

No. = number; LVMMI = left ventricular muscle mass index; EF = ejection fraction; LVH = left ventricular hypertrophy; P < 0.05 is shown as \*

**Conclusion:** Cardiovascular involvement in LSD in children is especially relevant in MPS. In FD in children, cardiac anomalies are rare. Especially in MPS, routine echocardiographic evaluation is necessary in children.

The figure (below) shows a parasternal long-axis view image of a 14 year boy with MPS type II (Hunter) with typical thickening of the mitral valve (arrow); LA = left atrium; LV = left ventricle



#### O10-1 Enlarging the genetic spectrum of cardiomyopathies. Preliminary results of the MyEstela Pediatric Cohort Study

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**Introduction:** The knowledge of the genetic origin of pediatric cardiomyopathies has increased over the last years. Dilated

cardiomyopathy (DCM) is the most common form, Hypertrophic cardiomyopathy (HCM) and Left ventricular noncompaction (LVNC) are commonly inherited as an autosomal dominant trait. The objective of this study is to describe the genetic origin of a cohort of pediatric patients with cardiomyopathy.

**Methods:** Prospective observational cohort study from June to December 2010 of children followed up in a familial cardiomyopathies outpatients clinic diagnosed with idiopathic cardiomyopathy. Demographics, family tree and genetic test for protein-coding sequences of sarcomere protein genes (MYH7, MYBPC3, TNNT3, TNNT2, TPM1, MYL2, MYL3, ACTC, and TNNT1) were performed.

**Results:** 36 patients (44,4% male) were included in the cohort. 26 genetic results have been received and 10 (38.5%) were positive. 20% (1/5) were positive in the group under 1 year of age, 28,6% (4/14) in the group between 1 and 10 years and 71,4% (5/7) in the group over 10 years. 40% of the positive results were male.

	Num patients	Gender	Age (years) Mean (range)
DCM	15	7M/8F	3,6 (0-14,4)
HCM	9	6M/3F	9,3 (0,2-16,5)
LVNC	12	3M/9F	8 (1,6-14,3)
Total	36	16M/20F	6,5 (0-16,5)
	Positive results	Gene type/ num positives	
DCM	2/13 (15,4%)	2/2	
HCM	5/6 (83,3%)	5/5	
LVNC	3/7 (42,9%)	1/3	
Total	10/26 (38,5%)	5/10	

The mutations were found in 5 of the 9 genes studied: 5 MYH7 (1 HCM, 1 DCM, 3 LVNC), 2 TNNT (1 HCM, 1 DCM), 1 ACTC (HCM), 1MyBPC3 (HCM), 1 MyL3 (HCM). 7 of them were novel mutations, not previously described in literature (3 HCM, 2 DCM, 2LVNC). At present 3 families have been studied, 1 being a de novo mutation and 2 an inherited mutation. During the study period two patients with HCM were implanted with a Cardioverter Defibrillator, two DCM died, one infant of sudden death and a new born with a prenatal hydrops diagnosis.

**Conclusions:** Mutations in cardiac sarcomere proteins are a common cause of cardiomyopathy in our initial cohort of children, more frequently in the oldest group and in those with HCM and LVNC. The description of high rate of novel mutations will improve the knowledge on pediatric cardiomyopathies.

#### O10-2 Diagnostic Evaluation in Pediatric Pulmonary Hypertension (PH) – Results from the First Pediatric Global Registry: Tracking Outcomes and Practice in Pediatric Pulmonary Hypertension (TOPP)

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**Introduction:** The TOPP registry (31 sites, 20 countries) was designed to provide current demographic, diagnostic, clinical and outcome data in pediatric PH. One primary objective was to describe the current work up/testing in pediatric practices to diagnose PH. Patients diagnosed with PH between 3 mos and 18 yrs were eligible for enrollment. Investigators reported ECG, chest X-ray (CXR), echocardiography (ECHO), and Holters were done and results as 'normal' or 'abnormal.'

**Methods and Results:** Between Jan 2008 and Feb 2010, 456 patients were enrolled. RHC confirmed PH in 435 (95%); in the remaining 5%, the diagnosis was made by pathology or echo. The majority of patients had ECGs, ECHOs and/or CXR performed: ECG (n = 430, 94%; 90% 'abnormal'), ECHO (n = 439, 96%; 99% 'abnormal') and CXR (n = 406, 89%; 80% 'abnormal'). None of the patients had normal results for all 3 of these tests, although 2 tests were normal in 3% and 1 was normal in 18%. Additional tests included Holter (n = 94, 21%; 56% 'abnormal'), routine laboratory studies (n = 411, 90%), BNP (n = 97, 21%) or NTproBNP (n = 95, 21%), cardiopulmonary exercise testing (n = 34, 7%, 91%  $\geq$  7 years), 6 minute walk test (n = 175, 38%), pulmonary function tests (PFT) (n = 122, 27%), overnight oxygen saturation/sleep study (n = 129, 28%), lung scan (n = 104, 23%, 41% 'abnormal'), pulmonary angiography (n = 198, 43%, 43% 'abnormal'), chest CT (n = 189, 41%, 74% 'abnormal'), magnetic resonance imaging (MRI) (n = 42, 9%, 83% 'abnormal') and lung biopsy (n = 21, 5%, 90% 'abnormal'). Exercise tests were performed most often in patients >7 yrs (p < 0.0001).

**Conclusions:** ECHO, ECG, and CXR were the non-invasive tests most commonly used to evaluate patients. At least one of these tests was abnormal in all patients with PH confirmed by RHC, suggesting that they can be used collectively to identify affected patients. A complete work-up was not performed in all patients. The low rate of exercise testing can be partly explained by the inability of children <7 years to perform reliable testing. It remains to be determined if this more limited approach is appropriate to discriminate different PH etiologies.

#### O10-3

##### **Does reversed apical or basal left ventricular rotation recover following medical therapy in children with dilated cardiomyopathy? A two-dimensional speckle tracking imaging study**

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**Objectives:** Reversed left ventricular (LV) rotation and loss of LV twist have been associated with a more advanced disease stage in adults with dilated cardiomyopathy (DCM). If standard heart failure therapy induce recovery of reversed LV twist mechanics in these patients is unknown. This study 1) assessed if reversed LV rotation mechanics can be identified in children with DCM, and 2) to study the effect of standard heart failure treatment on reversed LV twist during follow-up.

**Methods:** We performed a retrospective review of our echocardiographic database to identify all children (aged <18 years) with DCM seen at our institution from 2009 to 2010. Patients were included when reversed LV rotation was present before initiation of standard

heart failure therapy with diuretics, carvedilol, and angiotensin converting enzyme (ACE) inhibitor. Patients with muscular dystrophy, mitochondrial or metabolic disorders, and tachycardia-induced cardiomyopathy or poor image quality were excluded. LV twist was assessed using two-dimensional speckle tracking echocardiography. Normal LV twist demonstrates counterclockwise apical rotation and clockwise basal rotation as viewed from LV apex. **Results:** During this study period, 14 children presented with DCM. Reversed rotational mechanics were observed in 6/14 children (43%, all male, median age 16 years, range 2 to 18 years). Three different patterns of reversed rotation were identified: reversed apical rotation (n = 3), reversed basal rotation (n = 2), and mirror-image rotation (n = 1, counterclockwise basal and clockwise apical rotation).

All patients received standard heart failure therapy for a median of 10 months, range 2 weeks to 18 months. During this period, reversed rotation normalized in 2/6 patients. Heart failure deteriorated in 2 patients, requiring heart transplantation or cardiac resynchronization therapy, or both. In 2 other patients, LV twist remained reversed during follow-up. Only patients with reversed basal rotation demonstrated recovery of LV twist mechanics.

**Conclusion:** Reversed LV rotation with loss of LV twist can be identified in children with DCM using speckle tracking echocardiography. Reversed LV twist may normalize in a subset of patients following standard heart failure treatment. Lack of LV twist recovery may identify patients at high risk for adverse clinical outcome.

#### O10-4

##### **Clinical characteristics and outcome of heart transplant in adults with Congenital Heart Diseases. Subanalysis of Spanish Registry on Heart Transplantation**

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**Introduction:** In spite of recent medical and surgical advances, some patients with congenital heart diseases (CHD) need a heart transplant (HTx) as adults. Consequently, It has become necessary to know clinical characteristics of these HTx candidates.

**Objective:** To analyze outcome in the Spanish adult transplanted population with CHD and compare it with the most frequent causes of adult transplantation (ischemic heart disease [IHD] and idiopathic dilated cardiomyopathy [IDCM]) and among different subgroups of CHD.

**Materials and methods:** From May 1984 to December 2009, 6,048 patients were transplanted in Spain. Pediatric transplants (<16 years), combined transplants, reHTx, HTx performed in other centers without adult CHD cases, and HTx for heart diseases other than IDCM and IHD were excluded. Therefore, study population included 3,166 patients (IHD: 1 888; IDCM: 1,223; CHD: 55). Study subgroups: CHD transplants were classified according to pathophysiology into four groups: 1) Single ventricle with different

degrees of pulmonary stenosis (n: 18). 2) Single ventricle with cavopulmonary shunt surgery (Glenn/Fontan) (n:10). 3) Congenitally corrected transposition of great arteries or transposition of great arteries with Mustard/Senning surgery (n: 10). 4) CHDs with different degrees of right ventricle overload (tetralogy of Fallot, Ebstein's disease, transposition of great arteries with pulmonary stenosis and Rastelli patch, double outlet right ventricle) (n: 17).

**Results:** Significant differences were found between the clinical profile of patients with CHD and other groups: younger age, less need for inotropes and minor renal dysfunction. The analysis of survival showed some differences between groups (CHD vs IHD,  $p = 0.05$ ; CHD vs IDCM,  $p = 0.5$ ; IHD vs IDCM,  $p = 0.0001$ ). Early mortality was different between the different CHD subgroups (group 1: 19%, group 2: 40%, group 3: 0%, group 4: 29%;  $p < 0.001$ ). The probability of overall survival was higher in CHD group, despite its high early mortality.

**Conclusions:** Percentage of adult with CHD transplanted in Spain is low (1%). Long term survival is higher compared with groups with different indications for HTx (IHD and IDCM). Nevertheless, early mortality is high in operated or unoperated patients with single ventricle physiology. European registries with a larger number of patients are needed to better define results in this group of patients.

#### O10-5

##### **Recurrent Post-transplant lymphoproliferative disorder in paediatric heart transplant recipients**

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**Introduction:** Survival after paediatric heart transplantation (HTX) has improved significantly in the last decades. The long-term outcome is affected by morbidities mostly caused by required immunosuppressive therapy. Post-transplantation lymphoproliferative disorder belongs to the major cause of morbidity. We report about recurrent PTLD in paediatric heart transplant recipients medicated in our institution.

**Method:** We analyzed the time course of disease, Epstein-Barr Virus (EBV)-status, immunosuppression, involved organs and mortality.

**Results:** From 1988 till 2010, 156 patients were treated after paediatric heart HTX in our institution. Seventeen patients (11%) suffered from a malignancy. Seven of them (64%) had more than one episode. The group with recurrent PTLD developed the first malignancy on average seven years after HTX (range: 2–13 years). Mean time of recurrence was three years (range: 1–11 years). Most common symptoms were lymph node swelling in six of seven patients (85%). Only two patients had an extra nodal manifestation. Histology of the first malignancy was polymorphic (57%, four of seven) and Hodgkin-like lymphoma in two patients (29%). Second malignancy showed a change in histology: Six of seven patients (85%) had extra nodal manifestations; most common site was the gastrointestinal tract (70%, five of seven). Histology was monomorphic in four of seven cases (57%) and showed Hodgkin-like lymphoma in two cases. Most cases were of B-Cell origin and all patients were Epstein-Barr virus positive. Furthermore, all patients had a CNI-treatment since transplantation and an initial treatment with Azathioprin and were switched to a monotherapy after diagnosis. Treatment of malignancy consisted in CD-20-antibody therapy or chemotherapy (depending on histology). One patient died.

**Conclusion:** About 11% of paediatric heart-transplant recipients developed PTLD; a high percentage having a relapse. Malignancy was almost always of B-cell lineage and driven by Epstein-Barr virus. Because of high morbidity and mortality, strategies to improve the early diagnosis and the therapy for PTLD are needed.

#### O10-6

##### **Survival adjusted with age is similar in adults with Eisenmenger syndrome and other types of pulmonary artery hypertension**

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**Introduction:** Patients with Eisenmenger syndrome (ES) are considered to carry a better prognosis than other pulmonary artery hypertension (PAH) causes because of a more favourable hemodynamic profile. However, patients with ES usually present younger than other patients with PAH. Hence, such comparison for survival is potentially biased if this is not adjusted for age. Finally, the survival of patients with PAH consecutive to late closure of congenital heart defect (CHD-PAH) is not clarified. We aimed to compare the survival adjusted for age in ES, CHD-PAH and other causes of PAH.

**Methods:** We retrospectively studied all patients with PAH followed in our tertiary centre since 2003. Events outcome were defined as death or transplantation.

**Results:** Out of 150 patients with PAH, 32 had ES (21%) and 11 had CHD-PAH (7%). Median follow-up was 51 (26 to 81) months. The median age at presentation was lower for patients with ES and CHD-PAH: 26 (7 to 46) and 38 (18 to 56) years, respectively, versus 54 (41 to 66) years for patients with other causes of PAH, ( $p < 0.001$  and  $0.006$  for ES and CHD-PAH, respectively). Events during follow-up occurred in 8 (25%) of ES, 2 (18%) of CHD-PAH and 34.7% of patients with PAH. Patients with ES experienced significantly less events compared to patients with PAH (8 versus 48,  $p = 0.019$ ). Six patients with ES died, due to complication of cyanosis in 4 cases. The outcome for patients with CHD-PAH was similar than patients with other causes of PAH ( $p = 0.3$ ). Crude hazard ratio of events in ES and CHD-PAH population compared to patients with other causes of PAH was 0.3 (0.1 to 0.8) and 0.4 (0.1 to 1.8), respectively. When adjusted for age, ES and other causes of PAH experienced a similar longevity (0.6 range 0.2 to 1.9).

**Conclusion:** The longevity is similar between ES, PAH-CHD and other types of PAH. The long-term complications of cyanotic heart disease may counterbalance the relative protection of ES for right ventricular dysfunction.

#### O10-7

##### **The impact of cardiac resynchronization therapy on outcome of infants with severe congestive heart failure due to dilated cardiomyopathy – the importance of detecting “optimal pacing site” in OR**

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**Background:** The prognosis of dilated cardiomyopathy (DCM) with onset in infancy has been still dismal and poor, although cardiac resynchronization therapy (CRT) showed suboptimal improvements of some cases with electro-mechanical dyssynchrony.

**Aim:** To investigate the outcome of DCM with onset in infancy after CRT using epicardial leads and scrutinizing the search for optimal pacing site by intra-operative transthoracic (TTE) and transesophageal echocardiographic (TEE) monitoring using radial strain of speckle tracking imaging.

**Subjects:** 5 consecutive patients with DCM age at onset were ranged 2–6 months, and age at CRT ranged 9–13 mo, weight of 6–7 Kg. All 5 patients were in NYHA class IV and their left ventricular fractional shortening (LVFS) were 1–7% with



dyssynchrony (Ts-SD: 65–164 ms), BNP 1720–7680 pg/ml under catecholamine d.i.v.

**Method of CRT:** Because of small body size, we used a DDD pacemaker (ADDR01, Medotronics) with Y-shaped epicardial ventricular lead instead of InSyncIII. The optimal pacing site for LV epicardial lead was defined at both preoperatively and intra-operatively, as the latest contraction site of LV on radial and circumferential strain by TEE and TTE. Then RV pacing site was set with maximal trans-aortic flow velocity integral (VTI) and minimum dyssynchrony index. Finally the optimal atrio-ventricular delay time (AVDT) was set by maximum Ao-VTI and minimum QRS duration of ECG.

**Results:** 4 of 5 pts improved NYHA class from IV to I, but one was lost by sepsis. The 4 survivors improved LVFS from  $5 \pm 2\%$  to  $38 \pm 5\%$ , reduced BNP from  $4093 \pm 2890$  to  $15.8 \pm 6$  pg/ml. All dyssynchrony index (Ts-SD-TDI, Td-TDI, Ts-SR, Td-SR) were improved significantly all through the follow-up period (22–112 mo).

**In Conclusion:** CRT for infants with severe DCM, using epicardial lead, is feasible and promising treatment and could avoid transplantation, once one could assess the optimal pacing site appropriately and could set during operation by TTE or by TEE.

#### O11-1

##### **Prior administration of urinastatin decrease the total dosage of gamma-globulin in early stage Kawasaki disease**

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**Background:** High dose of intravenous gamma-globulin therapy (IVIG) is established treatment of Kawasaki disease (KD). However, there have been previous reports that IVIG in extremely-early-stage of KD made it probable that patients need additional therapies. On the other hand, urinastatin is known for one of the ancillary therapies of KD due to the mechanism of elastase inhibition.

**Objectives:** To evaluate the efficacy of the prior administration of urinastatin to KD in early stage.

**Subjects and Methods:** From January 2005 to March 2010, a total of 132 hospitalized patients who were diagnosed with KD within 4 days of the onset of symptoms were enrolled. The patients were separated into two groups. Group U (n = 25): who had administration of urinastatin prior to IVIG, Group N (n = 107): who had no administration of urinastatin prior to IVIG. Clinical data, clinical course, and circumstances of additional therapies were compared.

**Results:** Urinastatin significantly decreased fever in group U, compared to that in group N (38.5 degree C vs. 39.1 degree C,  $p < 0.05$ ). The duration of fever was shortened in group U compared to that in group N (6.0 days vs. 7.1 days,  $p < 0.05$ ). There were more patients who received 1g/kg IVIG for initial therapy was enough to improve symptoms in group U compared to that in group N (60.0% vs. 29.3%,  $p < 0.01$ ). There were fewer patients who received additional therapies when IVIG started on the 4th day of the onset of symptoms in group U compared to that in group N (20.0% vs. 35.6%,  $p < 0.05$ ). There were fewer patients who received additional therapies when the patients were two-day administration of urinastatin prior to IVIG on the 4th day of the onset of symptoms, compared to when the patients were one-day administration of urinastatin prior to IVIG on the 5th day of the onset of symptoms (0% vs. 66.7%,  $p < 0.05$ ).

**Conclusions:** Present study revealed that prior administration of urinastatin to Kawasaki disease which was diagnosed in early

stage offers relief of symptoms and decrease the amount of IVIG requirement. We suggest that this will lead to the practical reduction of healthcare cost.

#### O11-2

##### **Effect of dispensing with antibiotic prophylaxis before dental treatment in patients with congenital heart disease: Analysis of 21 consecutive cases of infective endocarditis after the introduction of NICE guidelines in April 2008**

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**Introduction:** In 2008 the National Institute of Clinical Medicine (NICE) in the UK concluded that routine prophylaxis with antibiotics was no longer needed in patients with congenital heart disease (CHD) undergoing dental treatment. Since 2008 we have advised against prophylactic antibiotic cover for all CHD patients undergoing dental treatment.

**Methods:** All patients presenting with endocarditis from April 2008 to November 2010 were identified and assessed for common risk factors including prior or remedial dental work and infecting organisms.

**Results:** All 21 patients seen at our institution with infective endocarditis were included in the study. The underlying diagnoses were: unoperated perimembranous ventricular septal defect (4), Tetralogy of Fallot (3), double outlet right ventricle (3), coarctation of aorta (3), aortic stenosis (2); pulmonary atresia with ventricular septal defect (1), pulmonary stenosis (1), hypoplastic left heart syndrome (1), truncus arteriosus (1), atrioventricular septal defect (1) and structurally normal heart (1). A bicuspid aortic valve was present in 3 patients. An implanted pulmonary valve homograft was present in 5 (24%) of the patients. Organisms detected were: oral pathogens in 11, staphylococcus aureus in 4, nasopharyngeal commensals in 2, miscellaneous organism in 3 and culture negative in 1. In the 11 (52%) patients with endocarditis due to an oral pathogen, in only 1 (9%) was this related temporally to preceding dental work (without antibiotic prophylaxis). One child had spontaneously shed deciduous teeth and one had removal of a fixed brace (antibiotic prophylaxis not previously recommended). In four (36%) patients, severe caries or dental abscesses were found after the episode of infective endocarditis was diagnosed.

**Conclusions:** There has not been a noticeable increase in dental related episodes of IE since the change in guidance for antibiotic prophylaxis, as was feared by many, in the short time that the new guidelines have been in place. The need for high levels of dental hygiene, however remain, and will need greater emphasis now that our patients no longer carry antibiotic prophylaxis cards and are therefore not reminded as frequently as in the past. More prolonged review is required to confirm the appropriateness of the new policy.

#### O11-3

##### **Univentricular Hearts in Denmark 1977–2009: Incidence and Survival**

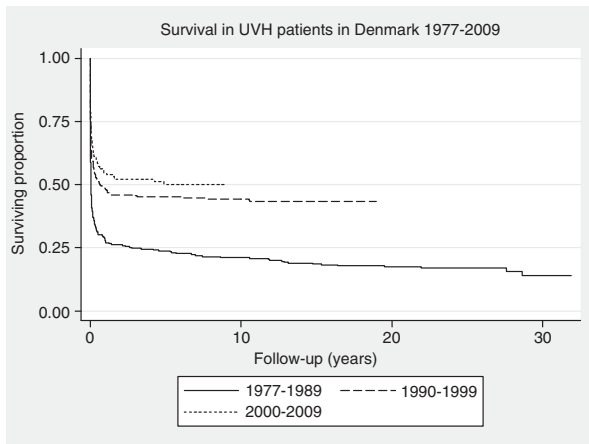
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**Introduction:** Over the past four decades outcome has improved in children born with a functionally univentricular heart (UVH). This study describes changes in incidence and survival among UVH patients born in Denmark from 1977 to 2009.

**Methods:** Using nationwide registries we identified UVH patients and induced abortions due to UVH in the fetus. The diagnosis

was confirmed through local surgical and fetal ultrasound registers, the national register of autopsy, and by review of medical and echocardiography reports. If UVH could not be confirmed the patient was excluded.



**Results:** 822 cases were identified. The incidence of UVH (including induced abortions) was evenly distributed in the period (mean 0.400 per 1000 births, SD 0.056). Incidence of liveborn UVH patients however showed a downward tendency in the period 2000–2009. Outcome of birth groups are shown below:

Birth group	Born with UVH, n			Fontan/TCPC, n (dead)
	Born with UVH, n	Abortion, n	Alive 2009, n	
1977–89	277	0	43	42 (10)
1990–99	248	6	106	99 (4)
2000–09	191	100	99	74 (4)

	Type of Fontan, n (dead)		
	1	2	3
1977–89	13 (4)	19 (6)	10 (0)
1990–99	1 (0)	66 (4)	32 (0)
2000–09	0	5 (2)	69 (2)

Fontan 1: Atriopulmonary connection; Fontan2: Lateral tunnel; Fontan 3: Extracardiac tunnel.

The Kaplan-Meier curves above (abortions excluded) show an increase in survival between 1977–89 and 1990–99 (log-rank,  $P < 0.0001$ ) and between 1990–99 and 2000–09 (although not statistically significant, log-rank,  $P = 0.20$ ). Survival and numbers of nondeceased/noncensored cases are shown in the table below.

Birthgroup	n	1 year survival (95% CI)		5 year survival (95% CI)		10 year survival (95% CI)			
		n		n		n			
1977–1989	277	0.28	(0.23–0.33)	76	0.24	(0.19–0.29)	65	0.21	(0.17–0.26)
1990–1999	248	0.48	(0.42–0.54)	121	0.45	(0.39–0.51)	113	0.44	(0.38–0.50)
2000–2009	191	0.54	(0.47–0.61)	93	0.50	(0.43–0.57)	49		

Birthgroup	n	20 year survival (95% CI)		30 year survival (95% CI)		
		n		n		
1977–1989	57	0.17	(0.13–0.22)	45	0.14	(0.09–0.20)
1990–1999	96					
2000–2009						

**Conclusions:** The incidence of UVH was constant throughout the period indicating constant detection rate over time. The incidence of liveborn UVH patients was falling after 2000 due to introduction of fetal ultrasound. Survival has improved from the 1980ies but has not significantly improved in the last 20 years.

#### O11-4

##### Pulse oximetry home monitoring in infants with single ventricle physiology and a surgical shunt as the only source of pulmonary blood flow

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**Introduction:** Thrombotic shunt occlusion is a major cause of death in children with single ventricle physiology and a surgical shunt as the only source of pulmonary flow. In a prospective non-randomized study we evaluated if home monitoring (HM) of oxygen saturation (SpO<sub>2</sub>) would detect life-threatening shunt complications between discharge after stage I and re-admission for stage II.

**Methods:** Twenty-four infants with single ventricle operated 2007–2010 with a BT (N = 7), central (N = 1) or Sano shunt (N = 16) were included. All were on aspirin 5 mg/kg/day and 3 on warfarin. Parents were instructed to measure SpO<sub>2</sub> once daily when the baby was calm and if SpO<sub>2</sub> < 71% repeat the measurement after 15 minutes. Positive HM was defined as SpO<sub>2</sub> < 71% on repeated measurement. By definition a shunt complication was present if there was arterial desaturation and narrowing of the shunt treated by intensified anticoagulation, surgical replacement, balloon dilatation, stenting, or with an earlier bidirectional cavopulmonary anastomosis (BDCPA). Parents' attitude towards the method was investigated using a questionnaire.

**Results:** A shunt complication occurred 11 times in 8 patients. HM was positive in 8 of 11 shunt complications (73%). In 2/8 shunt complications HM was probably life-saving; one patient had shunt endocarditis and the shunt was replaced the same day (age 59d), the other had an emergency balloon dilatation because of severe shunt narrowing (age 47d). In 3/11 shunt complications HM was negative; one of these 3 patients had an earlier BDCPA and survived but 2 died suddenly at home (age 42\* and 221d) from thrombotic shunt occlusion (autopsy), one of them\* only 3 hours after an SpO<sub>2</sub> reading of >90%.

On 5 occasions HM was positive but there was no shunt complication, albeit other problems in most.

HM with pulse oximetry was well accepted by the parents according to the results of the questionnaire.

Twenty of 21 survivors had a BDCPA (median 187d, range 95–377) with no mortality. One had a heart transplant instead of BDCPA.

**Conclusion:** HM of SpO<sub>2</sub> is well tolerated by caregivers and has the potential to detect some of the life-threatening shunt complications between stage I and II in infants with single ventricle physiology.

#### O11-5

##### Evaluation of the aortic morphology and function in adolescents after coarctation (CoA) repair using MRI: New details of a systemic vessels disease

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**Introduction:** Patients after CoA repair frequently have premature morbidity and mortality due to hypertension. Restenosis accounts for only a minority of these cases. Hypertension may

be also associated with intrinsic abnormalities of the aortic wall. Therefore, the aim was to evaluate aortic morphology and elastic wall properties in adolescents after CoA repair and to correlate these results with severity of restenosis and arterial hypertension, non-invasive pressure gradients, time and kind of surgical procedure, and aortic arch geometry.

**Methods:** Prospectively, 89 patients (age  $17 \pm 6,3$  y: end-to-end anastomosis (45), patch plastic (15), subclavian flap (9), others (20)) and 20 controls ( $18 \pm 4,92$  y) were examined using a 1.5 T whole-body MR scanner. 40% of the patients were surgically treated in 1st year of life. In addition to 3D-MR angiographies and phase-contrast flow measurements, 2D-CINE MRI were performed to assess the relative change in the aortic cross-sectional areas at the level of diaphragm to calculate aortic compliance (C).

**Results:** Two thirds of all patients showed systemic hypertension ( $> 90$ th perc. of age correlated value;  $54\% > 95$ th p.), but more than half of them had not a significant stenosis (defined as  $\geq 30\%$ ). C was lower in CoA than in control group ( $3.30 \pm 2.43$  vs.  $4.67 \pm 2.21$  [ $10^{-5} \text{Pa}^{-1} \text{m}^{-2}$ ];  $p = 0.024$ ). Significant differences were found also between hyper- and normotensive patients ( $2.61 \pm 1.60$  vs.  $4.11 \pm 2.95$ ;  $p = 0,01$ ), gothic and romanian arch geometry ( $2.64 \pm 1.58$  vs.  $3.78 \pm 2.81$ ;  $p = 0,027$ ). There was a good correlation between C and hypertension ( $r = 0.671$ ;  $p < 0.01$ ). On the other side no correlation could be shown between C (and hypertension) and time or kind of repair, restenosis or pressure gradients.

**Conclusion:** The decreased aortic compliance at level of diaphragm, the high rate of persistence of hypertension without restenosis and the clear independency of time and kind of surgical repair confirm the hypothesis that CoA may not be limited to the small area of isthmus but may be a widespread (systemic) vascular anomaly with impaired elastic wall properties. This may potentially contribute to the decision for surgical or interventional approach in patients with restenosis and hypertension.

#### O11-6

##### **Evaluation of aortic morphology and elastic properties in children and adolescents with Ullrich-Turner syndrome (UTS): High prevalence of aortic changes already in early life**

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**Objective:** Patients with UTS have an increased risk for progressive aortic root dilatation and dissection. Abnormal biophysical wall properties may be responsible for this. To avoid major cardiovascular events, it is important to recognize the high-risk patients early. Aim of this study was to evaluate morphology and elastic properties of the aorta for the first time only in children and adolescents with UTS.

**Methods:** In a prospective, cross-sectional study we examined 37 conscious girls with UTS without cardiac history, and all treated with growth hormone (median age (years):  $13.5$  [ $6.7-20.5$ ], median height SDS  $-2.21$  [ $-3.44--0.39$ ]), and 12 healthy controls ( $14$  [ $9-18$ ]) using a 1.5 Tesla whole-body MR scanner. Contrast-free 3D-MR angiographies were performed to assess aortic morphology. Diameters were measured at ascending (AAO) and descending aorta (DAO) and their ratio was calculated. BSA-corrected AAO-diameter is defined as aortic size index (ASI). MR protocol included 2D-CINE MRI to assess the relative change in aortic cross-sectional areas at the level of

diaphragm to calculate aortic compliance (C). ECHO and ECG were performed routinely.

**Results:** Changes of aortic morphology were found in 32% of the patients, while some had more than one alteration. Dilatation or prolongation was seen in 7 patients, 9 patients had an increased AAO/DAO ratio  $> 1.5$  and 8 patients had an increased ASI  $> 2 \text{ cm/m}^2$ , representing aortic dilatation. Bicuspid aortic valves were found in 5 patients. Compliance was similar in UTS patients and controls:  $C = 7.02 \pm 2.82$  vs.  $5.54 \pm 2.41$  [ $10^{-5} \text{Pa}^{-1} \text{m}^{-2}$ ]; t-test:  $p = 0.13$ . In routine-ECHO, in only 2 patients an AAO dilatation could be detected. 3 patients revealed a prolonged QT interval.

**Conclusion:** One third of the UTS patients had pathologic aortic findings including the aortic valve, one quarter already exhibits signs of aortic dilatation in early life, although the elastic wall properties seem to be normal yet (under growth hormone therapy). Monitoring of all these parameters may be relevant for evaluating disease progression and treatment options. Therefore, MRI helps to detect morphologic changes early and is more sensitive than ECHO but needs its validation in routine care to improve cardiac morbidity.

#### O11-7

##### **Acute Neonatal Hypertensive Crisis With Circulatory Failure: Be Aware Of The Wolf In Disguise**

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**Background:** Acute arterial hypertension with circulatory failure in the neonatal period is rare and poorly understood. Delay of correct treatment could lead to grave long-term consequences or even death.

**Methods:** Retrospective study from 2007–2010 at our tertiary Neonatal and Cardiology Centre; all patients presented in circulatory failure at an age  $< 14$  days; with arterial hypertension and secondary LV dysfunction without aortic malformation.

**Results:** Six cases were identified; 5 term patients (age  $7 \pm 2$  days, PMA  $40.0 \pm 1.6$ w, BW  $3650 \pm 562$  g) and 1 premature (D7, PMA 28 weeks, BW 860g). All patients presented with circulatory failure due to a renin-angiotensin-aldosterone system mediated hypertensive crisis. All needed respiratory support and had increased lactate levels (median  $2.25$  mmol/l, range  $1.4-11.0$ ). Renal function was normal in 2/6 cases. Cardiac findings: LV FS 27% (range  $10-30\%$ ); mild to moderate Aortic Regurgitation (AR) in all (grade 2/4 in 1); mild to moderate Mitral Regurgitation (MR) in all (grade MR  $> 2/4$  in 2); Tricuspid Regurgitation (TR) grade 2/4 in 3. LV thickness was normal in 3, mild hypertrophy in 3. One patient with severe dysfunction FS 17% had 2 large apical thrombi. At presentation 4 pts had hypertension, 1 hypotension, 1 normotensive. In 2 cases hypertension only became obvious after restoration of output.

Administration of IV milrinone was successful with rapid improvement of the clinical condition. High aldosterone (median  $2396$  ng/l) and renin (median  $23.5$   $\mu\text{g/l/hr}$ ) were present in all patients. LV and mitral function normalized in all; residual AR  $< 1/4$  in 2pts. One pt has an abnormal renal function; 2pts need anti-hypertensives.

Predisposing factors for renal thrombi were present in 2 pts with an arterial umbilical catheter and thrombus formation (1 with renal infarction).

**Conclusion:**

1. Early neonatal circulatory failure due to arterial hypertension is rare.
2. The combination of LV dysfunction and central aortic regurgitation in a structurally normal heart, should alert for arterial hypertensive crisis.



- Renin-angiotensin-aldosterone system mediated hypertensive crisis is most frequent.
- At presentation, hypotension (with dysfunctional LV) does not exclude a hypertensive crisis.
- Early recognition and urgent treatment is crucial.
- Cardiac long-term outcome appears good, renal outcome is variable.

#### PW1-1

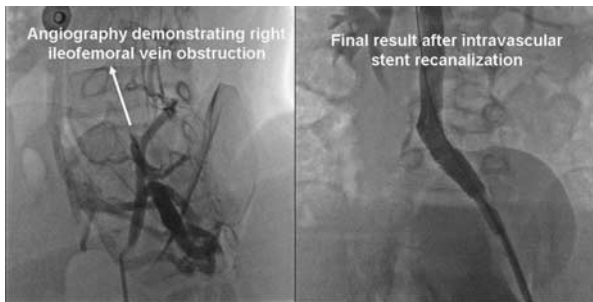
##### Stent recanalization of occluded iliofemoral and inferior cava veins in children with congenital heart disease: Acute and mid-term results

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**Introduction:** Patients with congenital heart disease present a high risk of systemic vascular damage following catheterizations and indwelling lines. The limitation of vascular accesses can be problematic in these patients, requiring multiple surgeries and/or diagnostic and interventional catheterizations in the future.

Our objectives are to evaluate the safety and efficacy of stentotic or occluded iliofemoral vein and inferior vena cava stent recanalization, and to determine the mid-term outcome following these procedures.

**Methods:** All patients with a vein occlusion and stent placed in the iliofemoral veins or in the inferior vena cava over a six year period (2004–2010) were included. Medical records and images were reviewed retrospectively in order to gather the following variables: age, weight, type and number of stents and follow-up procedures.



**Results:** 34 stents were placed in 17 patients (21 vessels). Mean age was 5,7 years (1,3–10,1) and weight 19,2 Kg (4,4–34). All patients had complex congenital heart disease of which five were heart transplanted patients and five had undergone a univentricular correction. 16 vessels were totally occluded, versus 4 subtotal stenosis. 2 patients had bilateral iliofemoral total occlusion, in these patients, the inferior vena cava was also occluded. The mean minimum vessel diameter in subtotal occlusion increased from 1,8 mm (1,1–2,5) to 8,8 mm (7,5–9,1). There were no major procedure complications. 4 patients (23%) underwent major surgery after recanalization. 9 patients (53%) had a total of 23 follow-up catheterizations, with a median length follow-up of 4 years. Two vessels (10,5%) were reoccluded requiring additional balloon dilation of existing stents. 11 patients were on chronic treatment with AAS and three patients had additional treatment with clopidogrel.

**Conclusions:** Stentotic or occluded vessel recanalization by intravascular stent placement is secure and effective to re-establish venous access for future catheterizations and/or surgeries. Reocclusion is unusual in our experience. Previously placed stents can be recanalized if necessary without major complications.

#### PW1-2

##### Double wire/double balloon/single stent delivery technique broadens interventional possibilities in challenging targets

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**Background:** Stent delivery can be difficult-dangerous in stenosis close to a bifurcation (jail-exclude vessels), or when external compression without anatomic stenosis (insufficient anchorage and risk of embolisation).

**Methods:** Prospective study tertiary interventional centre; all stenotic targets close to bifurcation where a double wire-double balloon-single stent technique was used.

**Results:** 6 patients:

A/Complex coarctation : n=2 (age 18 & 19y); large left subclavian artery originates just proximal of coarctation; through 14F sheath a 0.035" wire in ascending aorta & left subclavian artery; covered CP stent mounted on 4F & 16–18 mm Z-Med or BIB balloon; stent delivery & flaring with double balloon ; stent expansion with 20 mm Mullins balloon.

B/Pulmonary artery stenosis just proximal to bifurcation: n = 3 (age 2,3 & 7y); stent Genesis PG1910 over 2 balloons: 7 mm Opta +6 mm Maverick (0.014" Ironman) through 8F sheath, 7 + 7 mm Opta's through 10F sheath, 5 + 7 mm Opta's through 10F sheath. The stiff 0.014" Ironman wire gave poor support when compared to the 0.035" wires. After stent deployment, the balloons were advanced halfway out of the stent & reinflated, obtaining maximal flaring of the stent.

C/Pulmonary vein compression in Fontan candidate (20y); left common pulmonary vein "compressed" down to 3 mm slit-like passage by thoracic aorta and bulging atrial wall; no discrete stenosis by balloon inflation; balloon stretched diameter of 2 lobar veins determined; Genesis PG1920 on 10 & 12 mm balloon over 2\*0.035" wire through 12F sheath as distal as possible; opening of stent in "Y" resulted in adequate retention of stent. Enhanced runoff resulted in decreased PVR < Fontanable.

The desired result was obtained in all patients: the obstruction was relieved, all vascular pathways nicely open, good anchorage of the stent.

**Conclusion:** double wire-double balloon-single stent is a very useful technique in challenging targets close to a bifurcation. In order to keep sheath size limited, stent delivery & anchorage is done with double balloon, but stent expansion if needed with subsequent large (high pressure) balloon. Adequate support must be provided by wires: stiffer 0.035" wires are preferred in angulated situations (RVOT, PV).

#### PW1-3

##### Percutaneous Fenestration Closure using the Amplatzer Ductus Occluder II – Increasing the Experience

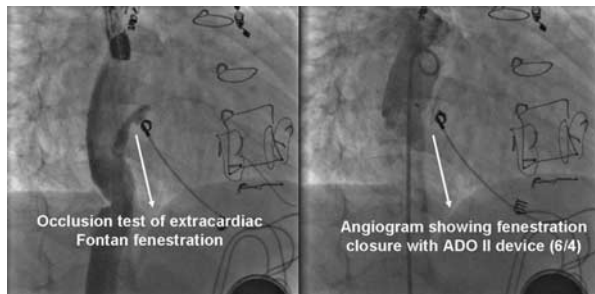
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**Introduction:** In the late 60's Fontan/Baudet described the basis for the surgery that became the final palliative surgery for single ventricular lesions. Since then, a relevant number of changes have been added to the classical Fontan operation, as Fontan baffles fenestration. With the aim of improving O<sub>2</sub> saturation and withdrawing anticoagulation, percutaneous fenestration closure in patients with a stabilized chronic hemodynamic situation

became a routine procedure in many centers. To this purpose, many different devices have been used.

**Aim:** Evaluate the efficacy and safety of the Amplatzer Duct Occluder II (ADOII) for Fontan fenestrations occlusion.

**Methods:** Retrospective review of medical records of all patients undergoing fenestration closure after Fontan procedure in the last 3 years. Each patient had complete right and left heart catheterization and fenestration test occlusion with a Thysak Mini Balloon, to assess hemodynamic suitability for closure (conduit pressure <15 mmHg); defect size was determined by transesophageal echocardiography.



**Results:** During this period, 27 Fontan fenestrations were percutaneously closed; in 22 cases (81.4%) ADOII was the implanted device (10 with extracardiac conduit and 12 with intracardiac lateral tunnel). Mean age was 7 yrs (18–5 yrs) and mean weight was 25Kg (18–81 kg). All the patients tolerated test occlusion and fenestration device occlusion without significant changes in arterial or venous pressures. Mean pressure in Fontan circuit increased from 10.4 mmHg (6–15 mmHg) to 11.7 mmHg (8–15 mmHg). PaO<sub>2</sub> showed a mean increase of 51.8 mmHg – range: 17–92 mmHg; FiO<sub>2</sub> <40% (p < 0.001). Only one patient presented significant device protrusion in the baffle, which was retrieved and replaced by a covered stent, with good result. Immediate post-implantation angiogram revealed trivial residual leaks in 11 patients, but subsequent follow-up echocardiography showed complete occlusion in all patients. The device position and integrity was satisfactory and there were no complications during the follow-up period.

**Conclusion:** ADOII device can be used safely and successfully in Fontan fenestrations percutaneous closure. Its low hemodynamic profile favours its easy implantation without significant protrusion on both sides of the conduit.

#### PW1-4

##### **Percutaneous pulmonary valve replacement in bovine jugular vein grafts (Contegra™) – Anything fancy?**

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**Objective:** Percutaneous pulmonary valve replacement (PPVR) is an established successful procedure for patients with degenerated surgically implanted grafts in pulmonary artery (PA) position. The bovine jugular vein graft (Contegra) as a new generation xenograft frequently does not show classical degeneration/calcification but the development of intimal proliferation at the anastomosis sites. Furthermore, one might presume that due to its elastic properties it might not be restrictive enough to accommodate the stented valve firmly and safely during PPVR. Therefore, the aim of our study was to compare the results of PPVR in patients with homografts and Contegras in PA position. **Method:** Retrospective comparison of the results of PPVR with

the Melody™ valve in paediatric patients with homografts and Contegras in our institution since March 2007.

**Results:** 13 patients with a homograft (mean age 12.7 (8–19) years), and 10 patients with a Contegra (age 12 (5–20) years) received PPVR. Patients with Contegra predominantly had stenotic lesions (n = 7), whereas homograft patients mostly had combined lesions with stenosis and regurgitation (n = 9). Contegras were treated earlier than homografts with PPVR – 5.5 (3.8–7.8) years vs. 11.3 (6.3–18.1) years after implantation (p < 0.01). There was no significant difference with regard to pre-interventional echo gradient (56 vs. 65 mmHg in Contegra). Apart from three all patients received pre-stenting of the graft with no differences in length of intervention and radiation-dosage. Contegra could be overstretched more effectively than the homograft (+2.6 mm to original diameter vs. 1.6 mm). In the homograft group one patient had a perforation of the LPA with the guide-wire, he needed surgery. Satisfying acute and midterm (follow-up 24.5 vs. 19.3 months in Contegra) gradient relief (>45%, p = 0.3) without any significant regurgitation could be achieved in both groups. In one homograft patient surgical explantation of the valve was performed 13 months after implantation because of a persisting systolic gradient >50 mmHg.

**Conclusions:** Contegras can be treated effectively with PPVR. It is even possible to overdilate them in comparison to their original diameter (mean +13%). Possibly also due to a modified surgical technique of side-to-side connections Contegras do have a roomier in- and outflow. Furthermore, this technique avoids distortion and kinking which results in straight grafts acting as a firm landing-zone for any percutaneously implanted stented valve.

#### PW1-5

##### **Growth of the Right Ventricle following an Aggressive Approach to Catheter Valvotomy in Pulmonary Atresia with Intact Ventricular Septum**

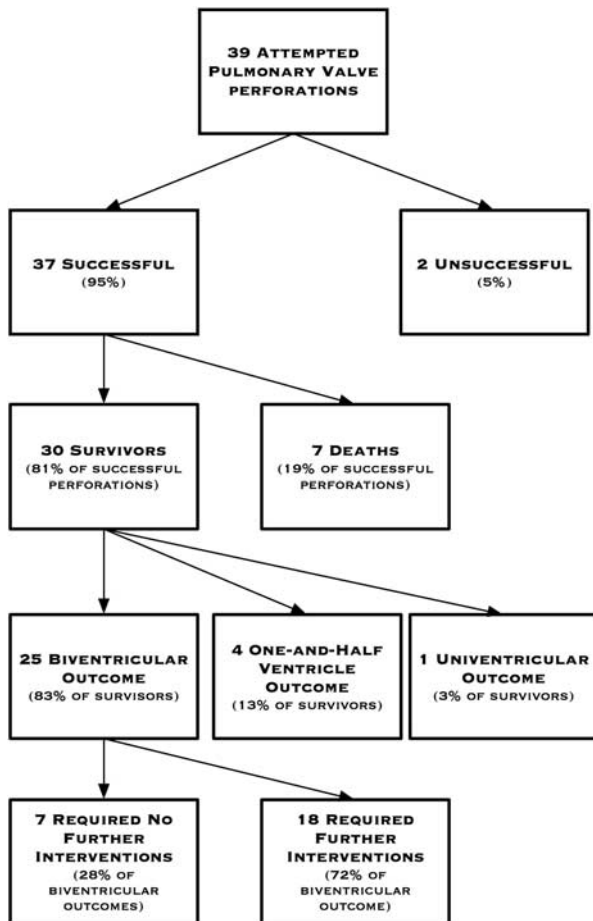
Chubb H., Pesonen E., Sivasankaran S., Simpson J.M., Krasemann T., Rosenthal E., Tibby S.M., Qureshi S.A.  
Evelina Children's Hospital, London, UK

**Introduction:** Catheter valvotomy of the pulmonary valve is now accepted as a common procedure for pulmonary atresia with intact ventricular septum (PAIVS). Since performing the first valve perforation in 1990, our centre has taken a relatively aggressive approach towards catheter valvotomy, performing the procedure in infants even with a very small right ventricle in an effort to establish forward flow and promote growth of the ventricle. We present the results of such an approach.

**Methods:** 39 infants underwent the procedure between 1990 and 2009, 37 successfully. The baseline patient characteristics were identified from the clinical notes and retrospective analysis of the initial echocardiograms. Long-term outcome was analysed at a median of 8.1 years (range 1–20 years) and included the current circulatory status and size of the tricuspid valve (TV). All available echocardiograms were reviewed, with a total of 141 sets of measurements obtained.

**Results:** There were seven (17%) deaths during the initial 35 days following successful valvotomy, and no late deaths occurred. Median initial TV z-score was –4.4 (range +2.2 to –15.0). At last follow-up, 25 (83%) of the survivors had a biventricular circulation, four (13%) a one-and-a-half ventricle circulation and one (3%) a Fontan circulation. Mean final saturations in subjects with and without biventricular circulation were 95.4 (±3.9)% and 95.2 (±1.1)% respectively.

Final TV z-score of patients achieving a biventricular outcome increased significantly ( $-4.4 (\pm 1.7)$  to  $-3.0 (\pm 2.0)$ ,  $p = 0.02$ ). TV z-score of those not achieving a biventricular outcome decreased significantly ( $-6.6 (\pm 1.8)$  to  $-10.1 (\pm 2.5)$ ,  $p = 0.02$ ).



**Conclusions:** This is the longest follow-up to date of survivors of this procedure, and there have been no late deaths. When there is no contraindication, extremely small right ventricles with small tricuspid valve diameters may be amenable to the procedure. With established forward flow, growth of the right ventricle may be achieved in some patients leading to a biventricular outcome.

#### PW1-6

##### **Percutaneous treatment of aortic isthmus atresia: use of radiofrequency perforation and covered stents**

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**Background:** Limited data exist in literature concerning the percutaneous treatment of aortic isthmus atresia.

**Patients and methods:** Between January 2007 and November 2010, 40 subjects underwent percutaneous treatment of aortic coarctation in our catheterization laboratory. Four out of 40 had complete aortic isthmus atresia. Median age at procedure was 48 years (range 32–63 years). All subjects had history of arterial

systemic hypertension refractory to medical treatment. Two subjects had a previous history of brain bleeding. All procedures were performed under general anesthesia and orotracheal intubation. Radial and femoral artery access were obtained. Radiofrequency system (Baylis MedComp Inc, Montreal, Canada) consisting in a Nykanen 0.024" RF guidewire and a coaxial microcatheter were used to perforate the atretic segment. Guidewire was snared and an artero-arterial circuit was created. The area was pre-dilated by using coronary angioplasty balloons. A 12 Fr Mullins long sheath was advanced and an E-PTFE covered 8Zig Cheatham-Platinum stent was implanted. Patients were monitored in hospital for 48–72 hours. Follow-up was performed at 1, 3, 6, 12 months and yearly thereafter.

**Results:** Percutaneous recanalization of the atresia was performed successfully in all subjects. Mean fluoroscopy and procedure times were  $30 \pm 6$  and  $90 \pm 15$  minutes, respectively. After implantation, the gradient decreased significantly (pre stent: mean value 52.25 mm Hg [range 33–70 mm Hg] versus post stent: mean value 3 mm Hg [range 0–10 mm Hg] [ $P < .0001$ ]). The stents were placed in the correct position in all subjects. No complications occurred. During a mean follow-up of 19 months (2–41 months), the results were stable without complications. All subjects had a perfectly normal arterial systemic pressure. In two out of 4 patient one anti-hypertensive drug was needed. One patient needed further stent dilation because of a conservative approach. Procedure was performed 8 months after the initial stent implantation without problem.

**Conclusions:** Our data show that use of radiofrequency energy and Covered CP stents is a safe, effective and promising tools for treatment of aortic isthmus atresia.

#### PW1-7

##### **Does presence of ventricular septal defect influence survival after arterial switch operation for transposition of the great arteries?**

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*Birmingham Children's Hospital, United Kingdom*

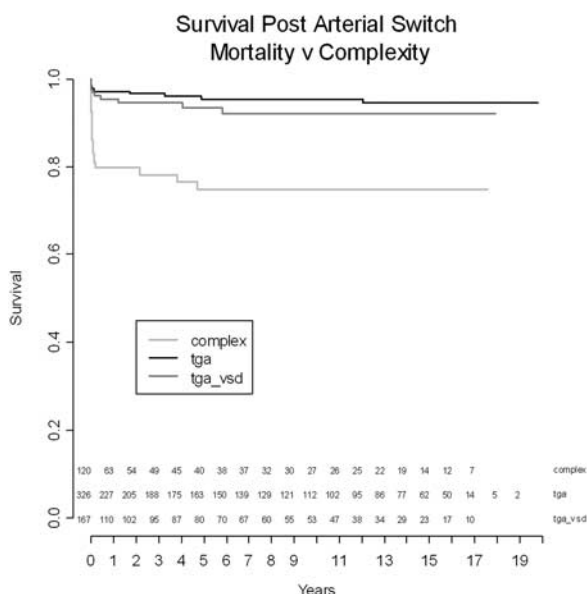
**Introduction:** The arterial switch operation (ASO) is well established as the basis for the operative treatment for infants with transposition of the great arteries (TGA). There is a broad range of morphological variation and therefore complexity of operative repair required for those infants with TGA. Risk scoring systems suggest operations for TGA with intact ventricular septum (TGA-IVS) attract a lower risk score than operations for TGA with ventricular septal defect (TGA-VSD) and operations for TGA with increasingly complex morphological variants. This long-term follow-up study was undertaken to examine the impact of complexity on survival after ASO.

**Methods:** Between 1988 and 2010, 613 infants (71% male, median age 8 days (IQ range 6–17 days)) underwent ASO for TGA at a single institution. For analysis, the patients were classified into three groups according to morphology, and therefore the surgical strategy used for repair: TGA-IVS ( $n = 326$ ), TGA-VSD ( $n = 167$ ), or TGA with complex anatomy i.e. other associated cardiovascular lesions (Complex,  $n = 120$ ). Infants with variants of TGA that did not undergo ASO were not included.

**Results:** Follow-up was 99.7% complete. Overall 30-day survival was 94.1%. Survival for those infants with TGA-IVS was 97.8% at 30 days, 95.5% at 5 years and 95.5% at 10 years. Survival for those infants with TGA-VSD was 96.9% at 30 days, 93.4% at 5 years and 92.1% at 10 years. Survival for those infants classified as Complex was 92.4% at 30 days, 74.7% at 5 years and 74.7% at 10 years. There was no



significant difference in survival between those infants with TGA-IVS and TGA-VSD ( $p = 0.3$ ), although those infants in the Complex group exhibited poorer survival than the other 2 groups (Figure 1,  $p < 0.001$ ).



**Conclusion:** Similar long-term survival can be achieved after ASO for TGA in those infants with intact ventricular septa or those with ventricular septal defects. However, infants with more complex anatomy continue to present a higher risk in the longer term.

#### PW1-8

##### Does neonatal cardiac surgery influence the balance between vasodilative and vasoconstrictive mediators?

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Department of Paediatric Cardiology (2); University Hospital Liège, Belgium

**Objectives:** Cardiac surgery in neonates is associated with the development a capillary leak syndrome (CLS), the exact cause of which remains unknown. The imbalance in the production of vasoactive mediators during and after surgery might be one of the key mechanisms in the pathophysiology of CLS.

**Aim:** To investigate the influence of neonatal cardiac surgery on the production of the vasodilative mediator nitric oxide (NO) and the vasoconstrictive mediator vasopressine as well as the postoperative outcome.

**Methods:** 27 neonates undergoing cardiac surgery were investigated. Plasma levels of nitrate and nitrite (as the end products of endogenous nitric oxide) and vasopressin were measured during and after surgery and correlated with clinical outcome.

**Results:** In all patients, a significant decrease in the plasma levels of nitrate/nitrite was observed during and after cardiac surgery. On the contrary, levels of vasopressin significantly increased during surgery and normalized on the first postoperative day. Duration of aortic clamping time correlated positively and esophageal  $T^{\circ}$  negatively with nitrate/nitrite concentrations. Lower nitrate/nitrite levels were associated with better cardiac function score and also with better renal function postoperatively. Higher vasopressin levels were associated with a better cardiac function and with an impaired renal function.

**Conclusion:** Neonatal cardiac surgery leads to decreased endogenous NO- and increased vasopressin production. This imbalance might correspond to a protective adaptation mechanism as it is associated with a better cardiovascular function postoperatively. Nevertheless, the fact that higher production of vasopressin is associated with impaired renal function might suggest that vasoactive imbalance in neonates contributes to fluid retention and therefore to the development of CLS in this patient population.

#### PW1-9

##### Propofol effect on cerebral oxygenation and cardiac output in children with congenital heart defects before cardiac catheterization

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**Introduction:** Propofol is a short-acting, intravenously administered hypnotic agent and is increasingly used in procedural sedation in children. However it decreases the sympathetic tonus and may lead to arterial hypotonia. “Electrical Velocimetry<sup>TM</sup>” offers the possibility for continuous non- invasive measurement of cardiac output. Near- Infrared Spectroscopy (NIRS) can measure cerebral tissue oxygenation in the frontal neocortex.

**Objective:** To evaluate the Propofol effect on cerebral oxygenation and cardiac output.

**Methods:** 31 children (f:m = 18:13), Median age: 49 (5–112) months, Median weight: 15,1 (4,8–34,3) kg. Continuous measurement of cerebral oxygenation and cardiac output was performed for 5 minutes before and after sedation with Propofol (1–2 mg/kg i.v.). Non invasive blood pressure and transcutaneous oxygen saturation (SpO<sub>2</sub>) was measured simultaneously.

**Results:** Propofol sedation led to a significant decrease in mean arterial pressure (MAP) and Cardiac index (CI). Cerebral tissue oxygenation index (TOI) however increased significantly ( $p < 0.05$ ).

Measurement	before Propofol	after Propofol
Heart rate (/min)	107.6 ± 24.3	110.0 ± 23.2
MAP (mmHg)	79.0 ± 15.5	66.7 ± 11.8*
SpO <sub>2</sub> (%)	92.5 ± 11.5	92.0 ± 9.5
TOI (%)	56.5 ± 10.6	58.7 ± 9.8*
Cardiac index (ml/min/qm)	3.2 ± 0.8	2.9 ± 0.7*

**Conclusion:** Propofol Sedation led to an increased cerebral tissue oxygenation despite a decrease in Cardiac index and arterial blood pressure. This may be caused by a decreased oxygen consumption of the sedated brain with intact cerebral auto regulation.

#### PW1-10

##### Fluctuations of Intraoperative Near Infra Red Spectroscopy During Neonatal and Infant Congenital Heart Surgery According to Defect Physiology, Bypass and Modified Ultrafiltration

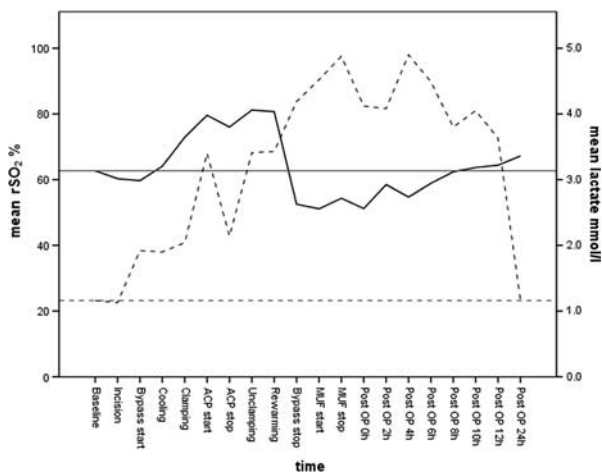
Gottschalk U. (2), Osse J. (1), Eulenburg C. (3), Goepfert M. (4), Wendt U. (2), Reichenspurner H. (1), Dodge-Khatami A. (1)

Division of Congenital Cardiovascular Surgery, University Heart Center (1); Pediatric Cardiology, University Heart Center (2); Institute of

Medical Biometrics and Epidemiology, University Hospital Eppendorf (3); Department of Anesthesia, University Hospital Eppendorf, Hamburg, Germany (4)

**Introduction:** Normal values of multisite Near InfraRed Spectroscopy (NIRS) at baseline, during, and after palliative or corrective congenital cardiac surgery are scantily defined, and may vary according to the defect and its corresponding physiology. We sought to analyze cardiac defect physiology-based fluctuations and the eventual effect of cardiopulmonary bypass and modified ultrafiltration on NIRS values.

**Methods:** 41 consecutive neonates and infants (19 biventricular acyanotic, 13 biventricular cyanotic, 9 single ventricle) undergoing protocol surgery with cardiopulmonary bypass were monitored with right + left cerebral and renal NIRS. Antegrade cerebral perfusion (ACP) was used in 10 patients. NIRS values, arterial lactate, and temperature at 20 time points were analyzed, from baseline 1 day pre-operatively, during bypass and modified ultrafiltration (MUF; 10 minutes), until 24 hours post-operatively. **Results:** In all patients, renal NIRS remained consistently higher than baseline, as did cerebral NIRS in all undergoing biventricular repair. During bypass (n = 41) and ACP (n = 10), there was no difference between right and left cerebral NIRS. Intra-operatively, cerebral and renal NIRS showed a weak inverse correlation with lactate levels (r = -0.17 and -0.155, respectively). Correlations between cerebral (r = -0.49) and renal (r = -0.101) NIRS and temperature were medium to weak, respectively. During MUF, reverse draining of oxygenated blood from the ascending aortic cannula and inflow perfusion through the right atrial cannula did not decrease cerebral or renal perfusion, and even slightly increased NIRS values (p < 0.001). In subgroup analysis of single ventricle patients (n = 9) and all undergoing ACP (n = 10; Figure 1), cerebral NIRS dropped below baseline at the end of bypass, and took respectively until 2 and 7 hours post-operatively to recuperate back to baseline values.



**Conclusions:** Different congenital cardiac defect physiologies have corresponding NIRS patterns at baseline, but are incompletely influenced during cardiopulmonary bypass. In patients with single ventricle physiology and those undergoing ACP, cerebral NIRS values lagged below baseline up to 2 and 7 hours post-operatively, respectively, although the long-term neurological relevance remains unknown. At bypass conclusion, ten minutes of MUF does not adversely affect cerebral or renal NIRS, and even increases both values. Bi-parietal cerebral NIRS monitoring is probably not warranted.

## PW1-11

### Home surveillance program clearly reduces interstage mortality after the Norwood operation for patients with Hypoplastic left heart syndrome

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**Objectives:** Since 1996 the hospital mortality after the Norwood operation of children with hypoplastic left heart syndrome (HLHS) in our centre could be steadily decreased to recently less than 3%. However, the interstage mortality till the superior cavopulmonary anastomosis (SCPA) remained high and was a major concern. Therefore we established a home surveillance program with the aim to overcome this problem.

**Patients and methods:** 45 infants with HLHS surviving the Norwood Operation between 10/05 and 09/09 were enrolled into the program. For comparison, infants before the start of the program (historical controls; n = 99) were used as well as 20 infants who were not discharged between the first and second stage operation. During the initial stay in the hospital parents were taught the handling of a monitor which recorded heart rate, respiration and arterial oxygen saturation. Additionally they were trained in cardiopulmonary resuscitation, recording daily saturation, heart rate, weight, fluid intake and instructed when to call the hospital (Saturation < 75%, no weight gain of 20 to 30 g in 3 days or an acute weight loss of more than 30 g in a day). An experienced paediatric cardiologist called them at home once a week.

**Results:** Interstage mortality was reduced significantly (p = 0.037) from 14.1% (n = 14/99) to 2.2% (n = 1/45). There was no significant differences between the three groups in their anatomical subgroups, the weight at the time of the Norwood operation or the shunt index diameter. At the time of SCPA children in the home surveillance program had a significantly lower age (102(67–299) vs 152 (77–1372) days) and weight (5.09 ± 0.79 vs 5.75 ± 1.22 kg) compared to those without it (p < 0.001). 14 infants did not meet the criteria all the time. 1 patient was late referred to the local hospital and died, 8 infants were operated earlier (SCPA n = 6, shunt replacement n = 2), but 5 could be discharged after observation.

**Conclusion:** The implementation of the home surveillance program led to a drastic decrease of interstage mortality. The time, effort and costs involved in such program are justified by the improved survival of this patient group. Therefore this should be continued.

## PW1-12

### Branch pulmonary artery flow reversal: Does it correlate with valve leak?

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The Children's Hospital, Denver, Colorado, United States

**Background:** The Contegra conduit is used for right ventricular outflow tract (RVOT) reconstruction in a variety of congenital cardiac diseases. We hypothesized that flow reversal in the branch pulmonary arteries may be secondary to compliance and increased capacitance in the Contegra conduit compared to normal human anatomy in young children, and not necessarily indicative of severe valvar insufficiency.

**Methods:** Retrospective chart review was performed for all patients less than 9 months of age who underwent RVOT reconstruction utilizing the Contegra conduit between March

2004 and August 2009 at The Children's Hospital in Denver, Colorado. Conduit position was classified as either anatomic or extra-anatomic. Post-operative echocardiograms were evaluated for presence of conduit insufficiency and branch pulmonary artery flow reversal.

**Results:** There were 152 patients who underwent right ventricular outflow tract reconstruction using the Contegra conduit between March 2004 and August 2009. Of the total number of patients, 59 were less than or equal to 9 months. Mean age was 83 days  $\pm$  84.72, and mean weight was 4.44 kg  $\pm$  1.16 kg. Thirty-seven patients were male. Twenty-six patients had the conduit implanted in an anatomic position, and the remaining 33 were extra-anatomic. Two patients had previously had a conduit, one being a Homograft and the other a Contegra. The 30 day mortality was 5% (3 patients). Post operative echocardiograms were available for review by a single cardiologist for 55 patients. Branch pulmonary artery flow reversal was seen in twenty echocardiograms, of which 10 had less than mild conduit insufficiency.

**Conclusion:** Flow reversal in the branch pulmonary arteries was not only seen with severe insufficiency of the Contegra valve. Thus, flow reversal by itself should not influence the diagnosis of conduit insufficiency or the indications for replacement. This phenomenon may be unique to younger patients secondary to the relative size of the conduit in relation to the patient, and capacitance of the conduit.

#### PW2-1

##### Acquired Right Ventricle Outflow Tract Obstruction in Twin-Twin Transfusion Syndrome

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**Introduction:** Twin-Twin Transfusion Syndrome (TTTS) complicates 4 to 26% of diamniotic monochorionic twin gestations. The majority of deaths associated with TTTS are due to cardiovascular compromise from congestive heart failure. Right ventricle outflow tract obstruction (RVOTO) may occur in the recipient twin in at least 9% of pregnancies complicated by TTTS. The etiology is not completely understood. RVOTO can potentially progress in utero and may worsen neonatal outcome. **Methods:** Retrospective review of all cases of TTTS treatment at our hospital between March 2008–November 2010. Demographic dates, fetal echocardiograms, perinatal and postnatal outcome and catheterization records were reviewed.

**Results:** Fifty-two pregnancies complicated by TTTS were identified. 6 recipient twins (11,53%) had RVOTO prenatal diagnosis. None of the donor twins had structural heart disease, and no major additional structural heart disease was identified in the recipient twins.

Gestational age range was 16–23 weeks at RVOTO diagnosis. 5/6 cases reviewed photocoagulation (83%).

Fetal echocardiography showed right ventricle hypertrophy, moderate to severe tricuspid regurgitation and dysplastic pulmonary valve in 5/6 cases. Retrograde ductus flow was documented in all cases.

3 of 6 cases (50%) progressed to more severe obstruction during gestation. Delivery was between 29–38 weeks and none was indicated for RVOTO. Weight was between 1380 to 2340 gr. Neonatal echocardiograms confirmed RVOTO in all cases and prostaglandin infusion was necessary in 5 of 6 cases (83%).

Percutaneous pulmonary valvuloplasty was performed 1 to 75 days after birth. Pericardial tamponade requiring surgical drainage complicated one procedure.

Muscular subvalvular obstruction was predominant in one case and another case had combined valvular and supra-valvular obstruction due to calcification of the main pulmonary artery and required surgery at 2 months of life.

**Conclusions:** RVOTO complicates 11,5% of TTTS pregnancies. Diagnosis and possibility of progression does not differ from structural RVOTO. None of our cases suffered fetal hemodynamic compromise and no neonatal deaths occurred despite all patients needing neonatal catheterization. The high incidence of duct dependency underlines the importance of prenatal diagnosis and highlights the indication of echocardiography evaluation in all pregnancies complicated by TTTS.

#### PW2-2

##### Outcome of Hypoplastic Left and Right Heart Syndrome (HLHS and HRHS) after antenatal diagnosis in South Wales over a seven year period

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**Objective:** To assess the rate of antenatal detection of HLHS and its outcome both in the antenatal and postnatal period. This review was undertaken to help improve counselling and provide better surgical survival information for expecting parents in the future.

**Methods:** All cases of Hypoplastic Left and Right Heart Syndrome detected antenatally between January 2002 and December 2008 were included in the study. Fetal medicine and fetal cardiac databases at a tertiary fetal cardiology centre as well as CARIS central database in Wales were utilised to carry out the review. The notes were carefully scrutinised to rule out any confounding variables.

**Results:** There were 55 cases of HLHS and 15 cases of HRHS in South Wales over this period.

Outcome of antenatal diagnosis:

	Total no.	Antenatally detected	Termination	Still birth	Fetal Loss	Born alive
HLHS	55	50	24	2	0	24
HRHS	15	14	5	1	1	7

Outcome of antenatally diagnosed live births:

	No. of Live births	Died without surgery	Received Surgery	Died following surgery	Alive
HLHS	24	1	23	5	18
HRHS	7	1	6	1	5

Hypoplastic conditions of the heart are often isolated conditions with a low rate of other system abnormalities (12 out of 55 for HLHS and 1 out of 15 for HRHS). The rate of chromosomal abnormality with HLHS is 10% (5 out of 50) and 6.7% (1 out of 15) with HRHS.

**Conclusion:** HLHS and HRHS are one of the most high-risk lesions in children with congenital heart disease. Antenatal detection rate for both these conditions has been satisfactory at over 90% in Wales. Termination rate remains high despite substantial improvement in survival after surgery. Post surgical survival stands at 78% in our series for HLHS with a maximum



follow up of eight years and 83% for HRHS with a maximum follow up of five years. Counselling is essential for parents to make an informed decision.

### PW2-3

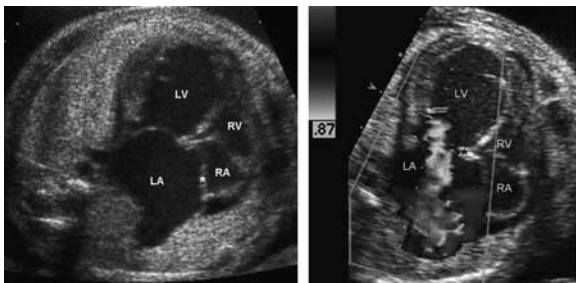
#### Aortic Stenosis and Severe Mitral Regurgitation in the Fetus Resulting in Giant Left Atrium and Hydrops: Pathophysiology, Outcomes, and Preliminary Experience with Prenatal Cardiac Intervention

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**Objectives:** To review anatomic, physiologic, and clinical features of fetuses and neonates with severe mitral regurgitation (MR) in conjunction with aortic stenosis (AS), and left ventricular (LV) and left atrial (LA) dilation, and present preliminary results of prenatal intervention for this condition.

**Background:** Severe fetal valvar AS with an abnormal mitral valve (MV) and MR can lead to left heart dilation, with consequent compression of the right ventricle (RV) (Figure 1a,b); hydrops and low cardiac output is often associated.



LV, left ventricle, LA, left atrium, RA, right atrium, RV, right ventricle, \*bright anterior mitral valve leaflet

**Methods:** Retrospective review of fetuses diagnosed with AS, severe MR, and LA dilation (2002–2009) and neonates with the same combination of abnormalities (1988–2009).

**Results:** Fourteen fetuses and 7 neonates were investigated. Eleven fetuses had severe hydrops; all had polyhydramnios and a structurally abnormal MV, abnormal MV inflow pattern, a restrictive/intact atrial septum, retrograde flow in the transverse aortic arch, and compression of the right heart. The mean indexed RV output was  $326 \pm 160$  mL/kg/min, substantially lower than the normal average fetal combined ventricular output of  $550 \pm 150$  mL/kg/min. Ten fetuses underwent prenatal cardiac intervention: aortic valvuloplasty ( $n = 8$ ) and/or atrial septal dilation/stenting ( $n = 5$ ). Seven of these, and 11 overall, were liveborn. Nine patients died (median age 6 days), 2 patients are currently alive. All 7 patients diagnosed in the neonatal period died (median age 1 day).

**Conclusions:** AS associated with significant MR in the fetus can cause severe LA enlargement, leading to low cardiac output and hydrops. Despite the potential advantages of early prenatal diagnosis and both fetal and neonatal interventions, this rare complex of anomalies carries a poor prognosis.

### PW2-4

#### Natural course of right ventricular dilation in severe pulmonary regurgitation after repair of tetralogy of Fallot

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**Introduction:** Tetralogy of Fallot (TOF) can be surgically repaired in the first months of life with low mortality. During long-term follow up, severe pulmonary regurgitation (PR) leads to right ventricular (RV) dilatation, impaired RV function and potentially lethal arrhythmias. Timely pulmonary valve replacement (PVR) may prevent these sequelae. However, the correct timing for performing PVR remains an open debate.

We sought to assess the rate of progression of RV dilatation in presence of severe PR.

**Methods:** 34 patients after TOF repair with PR and severe RV dilatation, age  $12.4 \pm 5$  y, weight  $45.8 \pm 18.3$  kg, underwent at least two consecutive MR examinations for evaluation of RV volume and function. The time interval since surgical repair was 11y (4–23). Time interval between first and last MR scan was 3y (1.1–6).

Ventricular volumes were calculated on a stack of short-axis images covering both ventricles acquired with the steady-state free precession sequence. A difference of  $>8\%$  between two measurements was considered significant.

**Results:** Initial RV dilation was more severe in patients after transannular patch ( $RVEDV 171 \pm 32$  ml/m<sup>2</sup>) than in others ( $145 \pm 18$  ml/m<sup>2</sup>) ( $p < 0.05$ ). No significant changes in volume and function occurred in the overall patient's group (table).

**Methods:** RV volume enlarged in 13/34 patients, remained unchanged in 11, and decreased in 10. In 14 patients with severe RV dilatation ( $RVEDV > 150$  ml/m<sup>2</sup>),  $RVEDV$  increased in 5, remained unchanged in 6 and decreased in 3. The overall mean rate of volume change was  $2 \pm 7$  ml/m<sup>2</sup>/year for the left ventricle and  $0.1 \pm 12$  ml/m<sup>2</sup>/year for the RV, without correlation with the technique of repair or the degree of dilatation.

**Conclusions:** During a period of 3 years progressive RV dilatation occurred in 38% of patients with severe PR. Increase rate did not correlated with the technique of repair, or the degree of RV dilatation. Analysis of different risk factors may help in properly selecting patients for PVR.

	MR 1	MR 2
RV EDV (ml/m <sup>2</sup> )	149 ( $\pm 35$ )	152 ( $\pm 38$ )
RV ESV (ml/m <sup>2</sup> )	78 ( $\pm 35$ )	81 ( $\pm 29$ )
RV EF%	48 ( $\pm 7$ )	48 ( $\pm 8$ )
LV EDV (ml/m <sup>2</sup> )	73 ( $\pm 13$ )	75 ( $\pm 13$ )
LV ESV (ml/m <sup>2</sup> )	34 ( $\pm 7$ )	34 ( $\pm 9$ )
LV EF %	55 ( $\pm 6$ )	56 ( $\pm 7$ )

### PW2-5

#### Endothelial function in Kawasaki patients

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**Background:** Kawasaki disease (KD) consists of an acute systemic vasculitis of unknown aetiology. Cardiac complications are

frequent and include endothelial dysfunction (ED) in patients with coronary anomalies. Up to now it has not been clearly demonstrated the presence of ED in KD patients who had no coronary lesions. Peripheral arterial tonometry (Endo-PAT) measures the reactive hyperemia mediated by NO release in response to local ischemia. It has been validated in adult population to access microvascular function, but its use in pediatric patients is scarce.

**Aim:** To evaluate endothelial dysfunction (ED) in children and young adults as long term complication after KD, using Endo-PAT.

**Methods:** Case-control study. Group A: KD patients aged more than 11, with KD diagnosed for more than 5 years and no coronary lesions or any other identified risk factor for cardiovascular disease including normal body surface mass. Control Group: individuals without cardiovascular risk factors, matched for gender and age. Patients and controls were accessed clinically, and by electrocardiography and echocardiography. Endo-PAT was performed to determine reactive hyperemia index (RHI) and augmentation index (AI). It was repeated one to three months later, to access consistency of data.

**Results:** 35 individuals were evaluated (Group A: 19 vs Controls: 16). Groups were comparable in terms of gender distribution (women: 37% vs 43%), age ( $21 \pm 6$  yo) and height ( $166 \pm 9$ ). Compared with control group KD patients presented significant lower RHI ( $1.68 \pm 0.49$  vs  $2.31 \pm 0.53$ ;  $p = 0.001$ , Mann Whitney test). AI was similar in both groups ( $-10 \pm 7$  vs  $-11 \pm 5$ ;  $p > 0.005$ ). The majority of patients with KD disclosed ED (68%) detected by RHI compared with 12% in controls. In fact KD patients had 5.5 folds increased risk for ED as a long term follow up complication of KD compared to controls.

**Conclusion:** Peripheral arterial tonometry is feasible, safe and reproducible in pediatric population to study endothelial function. Endothelial dysfunction is a frequent long term complication in patients after Kawasaki disease and normal appearing coronary arteries compared with healthy controls. However, these results need validation in a larger population.

## PW2-6

### Echocardiographic Assessment of Right Ventricular Volumes: A Comparison of Different Techniques in Children after Surgical Repair for Tetralogy of Fallot

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**Background:** Different echocardiographic techniques are available for assessing right ventricular (RV) volumes but their clinical validity has not been well established. We tested the feasibility, reproducibility and accuracy of 3 different echocardiographic techniques compared to MRI to measure RV volumes and ejection fraction (EF) in patients after tetralogy of Fallot (TOF) repair.

**Methods:** Forty patients (age  $13 \pm 2.9$  y) after TOF repair were studied using 3D volume acquisition analysis (3D) (Tomtec, Germany), 2D echo with knowledge based 3D reconstruction (KBR) (Ventripoint, USA) and the 4 chamber area (4C area) method. Parameters analyzed were end diastolic volume (EDV), end systolic volume (ESV) and EF. Ultrasound images were acquired using a Vivid 7 scanner (GE Ultrasound, USA). In 20 patients echocardiography was performed immediately after

cardiac MRI. Intra and inter-observer as well as inter-technique variability was assessed using Pearson correlation analysis (R), coefficient of variance (COV) and Bland-Altman analysis.

**Results:** Feasibility was 90% for 3D, 97.5% for KBR and 100% for 4C area method. Intra- and inter-observer variability were good for both KBR and 3D echo while more variability was observed for the 4C method (e.g. EDV R 0.997, 0.995 and 0.985; COV 3.2, 3.8 and 5.4; bias  $1.1 \pm 7.6$ ,  $-2.2 \pm 8.8$  and  $4.3 \pm 12.5$  respectively). The results for intertechnique variability are summarized in the table. Compared to MRI volumes KBR underestimated EDV by around 4% with narrow limits of agreement, while the 3D method underestimated EDV by around 10% with slightly wider limits of agreement. The 4C area method overestimated the volumes by around 10% with poor agreement. For the ESV, the inter-technique variability of KBR and 3D was slightly higher, with higher coefficients of variation and wider limits of agreement.

**Conclusions:** Current echocardiographic techniques to assess RV volumes are highly feasible in a pediatric population of TOF patients with the highest feasibility for 2D echo methods. KBR was shown to be the most accurate technique when compared to MRI with a small bias and relatively narrow limits of agreement. This study shows that echocardiography is approaching the reproducibility and accuracy of MRI-based RV volume quantification.

Inter-method variability	N	Correlation coefficient	Coefficient of variance	Mean bias $\pm$ SD	p
KBR versus MRI	20				
EDV		.992	4.1	$-4.8 \pm 9.8$	.04
ESV		.974	7.6	$-8 \pm 10.1$	.002
EF		.896	7	$2.1 \pm 3.2$	.007
3D versus MRI	19				
EDV		.983	6.2	$-10.1 \pm 14.4$	.006
ESV		.962	9.4	$-4.9 \pm 12.5$	.1
EF		.760	10.9	$-0.4 \pm 4.7$	.7
4C area versus MRI	20				
EDV		.967	7.8	$9.7 \pm 19.5$	.037

## PW2-7

### Ventricular flow dynamics in hypoplastic left heart: useful insights from patient specific computer modelling

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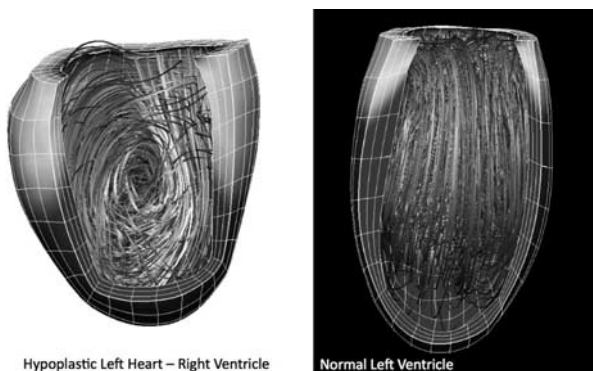
**Introduction:** The systemic right ventricle (RV) in hypoplastic left heart syndrome (HLHS) is significantly geometrically different from the normal systemic left ventricle (LV) or the systemic RV in a biventricular circulation. Computer modelling allows the construction of a patient specific dynamic heart model allowing visualisation of blood flow patterns in the ventricle.

**Methods:** Ethical and institutional approval was obtained. Echocardiography was performed under the same anaesthetic as cardiac magnetic resonance imaging (MRI). MRI derived geometries, tissue Doppler, pulsed wave Doppler and speckle tracking were combined with computational tools to produce a patient specific model. The interaction between blood flow and myocardial behaviour was investigated by numerical simulations highlighting the vortex formation mechanism and corresponding energy transfers inside the ventricle. Models from a patient with HLHS were compared to a normal systemic LV.

**Results:** In diastole the main fluid dynamic feature is the formation of ring vortices inside the ventricle. In the normal ventricle two ring vortices are generated, corresponding to

the E- and A-waves. This leads to two peaks of kinetic energy and to a reduced rate of viscous dissipation, reflecting the optimal energy configuration during the formation mechanism (where nearly all the inflow is entrained into the ring vortex). As each vortex shifts towards the apical region pressure waves are generated and the vortex contributes to myocardial displacement. In the dilated RV in HLHS, only one ring vortex is formed in the cavity centre with restricted axial displacement compared to the normal LV. This is associated with reduced pressure gradients and limited apical displacement and velocities, which correlated with the echocardiographic measurements. The higher rate of viscous energy loss in these patients could be linked to the lower energy efficiency of the single vortex, which is unable to absorb additional energy from the inflow and gives rise to a series of secondary swirling structures.

**Conclusions:** Patient specific modeling allows insights into the flow patterns within the ventricle and may therefore be helpful in understanding why the systemic RV does not perform as well as a systemic LV. Validation with haemodynamic data acquired during MRI catheter procedures is underway.



#### PW2-8

##### Right ventricular systolic function after Tetralogy of Fallot repair revisited: Does the outflow tract matter?

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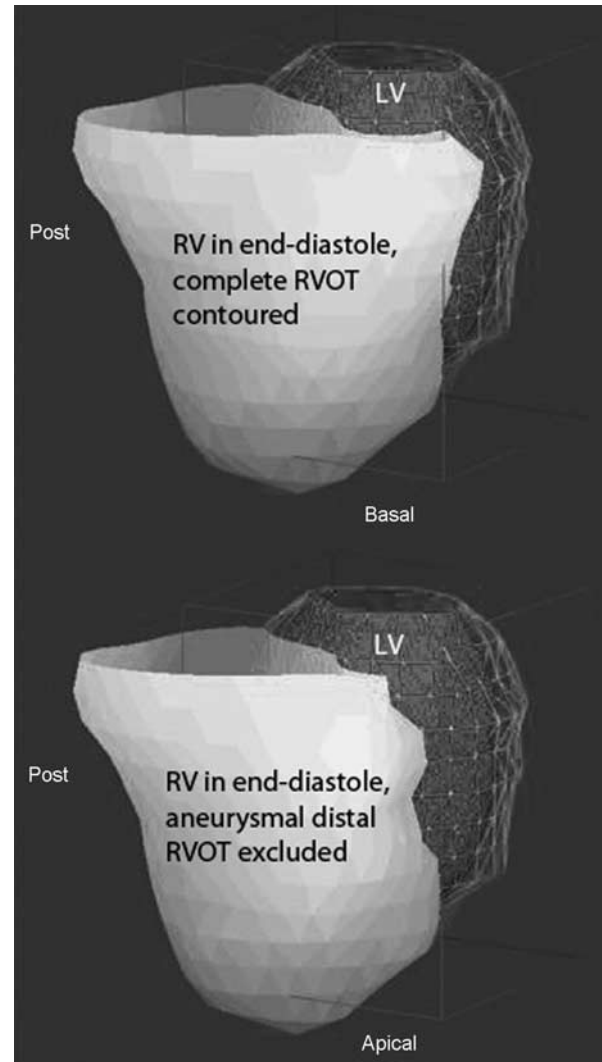
**Objectives:** Quantification of right ventricular (RV) systolic function following Tetralogy of Fallot (TOF) repair remains challenging due to the complexity of the RV geometry. Cardiac magnetic resonance imaging (CMRI) is the gold standard for assessing RV volumes. Nevertheless, controversy persists over whether to include the non-contractile RV outflow tract (RVOT) in the calculation of RV ejection fraction (EF, see Figure).

We aimed at evaluating various MRI and echocardiography derived descriptors of RV systolic function. In particular, we sought to assess the effect of excluding the aneurysmal RVOT after TOF repair on RVEF and to determine which of the examined methods serves as a better predictor of clinical status.

**Methods:** We reviewed the CMRIs, echocardiograms, and medical records of 50 consecutive patients with repaired TOF who were referred for CMRI. Cardiopulmonary exercise test results were used as surrogate parameters for clinical status. In addition to the routine CMRI data RV volumetry was repeated excluding the non-contractile RVOT in systole and diastole (Figure). The displacements of the RV inferior and lateral annulus were measured on CMRI.

**Results:** After excluding the akinetic RVOT, RVEF was higher ( $p < 0.0001$ ) and correlated slightly better with maximum oxygen consumption ( $VO_{2maxpred}$ ). ( $r = 0.36/p = 0.03$  vs.

$r = 0.33/p = 0.04$ ). RV inferior and lateral annular displacements correlated with  $VO_{2maxpred}$  ( $r = 0.49/p = 0.003$ , vs.  $r = 0.47/p = 0.049$ , respectively). Left ventricular EF, as measured by CMRI, RV end-diastolic volume and QRS duration were not predictive of exercise tolerance. Echocardiographically, RV wall longitudinal strain showed the best correlation with  $VO_{2maxpred}$ . The correlations of overall RV strain, average septal and lateral wall strains with exercise tolerance were  $r = 0.56/p = 0.0006$ ,  $r = 0.49/p = 0.003$  and  $r = 0.56/p = 0.0007$ , respectively.



**Conclusions:** Exclusion of the aneurysmal part of the RVOT in patients with TOF increased RVEF significantly, with a better correlation with exercise tolerance. However, a simple and quick MRI measurement – the displacement of the RV annulus in systole – showed a better correlation with  $VO_{2maxpred}$  than EF. RV wall longitudinal strain is the best echocardiographic predictor measures to predict exercise performance after TOF repair and LVEF does not determine exercise tolerance.

#### PW2-9

##### Development and validation of a novel automated learning based algorithm for quantification of MRI right ventricular volume in Tetralogy of Fallot

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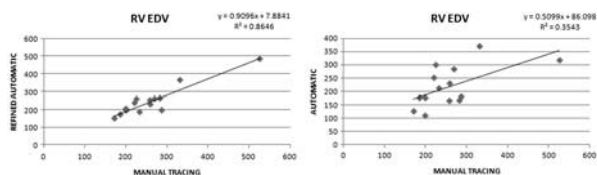


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**Introduction or Basis or Objectives:** Quantification of right ventricular volume (RVV) and ejection fraction (EF) in patients with complex congenital heart disease is essential and currently requires manual time consuming endocardial tracing. We compare the accuracy and post processing time of a novel automated learning-based algorithm (LBA) with current methods.

**Methods:** LBA relies on robust machine learning methods to capture complex statistics of right ventricular shape and appearance in cine steady-state free precession or gradient images. The LBA, previously trained on 114 patients with Tetralogy of Fallot (TOF), is applied to automatically estimate the three-dimensional RV boundary from images of unseen patients. To validate the LBA method, we examined 15 additional patients with TOF and compared three methods blindly in measuring RVV and EF: current standard manual tracing (M-TRACE), fully automated LBA (AUTO), and LBA with manual boundary refinement of fully automated tracings (R-AUTO). Using linear regression, right ventricular systolic (RVESV) and diastolic volumes (RVEDV) and EF were compared between manual and automated methods. Average processing time per patient was compared between techniques.

**Results:** Mean processing time (minutes) per patient was significantly reduced using the fully automated method but not when adding the overhead of manual correction: M-Trace,  $5.7 \pm 0.9$ ; AUTO, 0.7 ( $p < 0.001$ ); and R-AUTO,  $16.2 \pm 2.0$  ( $p < 0.002$ ). Results from manual tracing demonstrated poor correlation with diastolic volumes (RVEDV AUTO 0.595; R-AUTO, 0.930); good correlation with systolic volumes (RVESV AUTO, 0.874; R-AUTO, 0.918), and superior EF measures (AUTO, 0.854; R-AUTO, 0.633). LBA volume inaccuracy was most commonly due to incorrect identification of the tricuspid and pulmonary annuli. The figure below displays the relationship of RVEDV as measured by manual tracing, with the results from fully automatic and refined automatic techniques.



**Conclusions:** Our fully automated LDA method markedly reduces the time necessary to complete volumetric assessment of right ventricular function but due to failure to delineate accurately the tricuspid and pulmonary valve annuli, underestimates predominantly diastolic right ventricular volume. With additional case training, LBDA offers the opportunity to develop fully automatic volumetric measures to overcome the complexity of right ventricular function assessment of congenital heart disease and thereby reduce operator variability.

#### PW2-10

##### New Steady State Submaximal Exercise Test during Cardiovascular Magnetic Resonance – First Results in Healthy Volunteers

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Cardiovascular Magnetic Resonance (CMR) is important for assessment of cardiac anatomy and function. It lacks, however, the opportunity of performing exercise during scanning. Therefore, we established a new method of performing steady state submaximal exercise during CMR scans.

**Methods:** Ten healthy volunteers (median age 25 yrs., range 19–38) were studied by CMR and bicycle cardiopulmonary exercise test (CPET). By CMR, left ventricular (LV) volumes and stroke volume in the ascending aorta were measured under rest and submaximal exercise. Heart rate (HR), systolic (RRsys) and diastolic (RRdia) blood pressure were assessed, too. Steady state submaximal exercise was defined as 144 up and down strokes of the extended legs per minute, directed by an electronic metronome. For this purpose, a simple pulley was fixed to a specially designed frame and mounted on the MR table. The volunteers' legs were connected by a rope, passing over the pulley. CPET was performed at a standard symptom-limited bicycle exercise test and a second time using the steady state submaximal exercise setting from the CMR unit. During both tests oxygen uptake ( $VO_2$ ) and cardiopulmonary response were assessed.

**Results:** Maximal  $VO_2$  at routine CPET was  $40 \pm 7$  ml/kg/min. During steady state submaximal exercise using the CMR exercise setting the  $VO_2$  was  $9 \pm 2$  ml/kg/min. Thus the new exercise mode resulted in a mean stress level of  $24 \pm 5\%$  of maximal stress. During steady state submaximal exercise HR, RRsys and RRdia increased by  $33 \pm 10\%$ ,  $14 \pm 7\%$  and  $19 \pm 10\%$ , respectively.

LV enddiastolic and endsystolic volume decreased by  $5 \pm 12$  and  $14 \pm 18\%$ , respectively. LV ejection fraction rose by  $4 \pm 7\%$ . Cardiac index, assessed by phase contrast measurement, increased by  $30 \pm 10\%$ .

**Conclusion:** This new exercise mode enables steady state submaximal exercise in the CMR magnet bore, resulting in a mean stress level of 24% of maximal exercise. It enables composite hemodynamic measurements under stress with acceptable motion artefacts, that has not been feasible before.

#### PW2-11

##### Safety and Clinical Utility of Cardiovascular Magnetic Resonance in Neonates with Congenital Heart Disease

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**Introduction:** Cardiovascular magnetic resonance (CMR) is an established advanced diagnostic modality in adult with congenital heart disease (CHD). CMR is being increasingly used also in young children with complex CHD, as complement to echocardiography. We sought to evaluate the indications, safety and clinical impact of CMR performed in neonates with CHD.

**Methods:** Clinical records and imaging reports of all patients younger than 1 month, undergoing CMR between 2002 and 2010 were retrospectively reviewed for diagnosis, indication for CMR, complications and impact on clinical management. In all cases CMR was performed as second-line diagnostic examination complementing echocardiography.

**Results:** Seventy-eight CMR examinations were performed in 77 neonates. Mean age was  $7 \pm 8$  days, weight  $3064 \pm 633$  g.

Diagnosis included aortic arch anomalies in 23 children, pulmonary atresia/multicentric lung perfusion in 16, complex CHD with single ventricle in 13, complex CHD with two ventricles in 10, pulmonary vein anomalies in 8, tetralogy of Fallot in 2, tumour in 2 and 4 others. Correspondingly main indication for CMR was assessment of the aorta in 26 cases, pulmonary arteries in 21, pulmonary veins in 15, complex congenital heart disease in 8, myocardium in 3, ventricular size in 3, and two others. Mean scanning time was  $30 \pm 12$  min. The neonatal intensive care team performed anaesthesia with mechanical ventilation in 57 cases, anaesthesiology staff in 21. No significant complications occurred during examination. In two patients in critical condition breath-holding was avoided and the images acquired during free breathing. CMR findings had a major impact on further clinical management in 67/77 (87%) of the patients. The information obtained was crucial for following cardiac surgery in 54 cases, for catheter-guided intervention in 4; palliative care was decided in 9 neonates. In 7 children suspected diagnosis was confirmed and in other 4 ruled out.

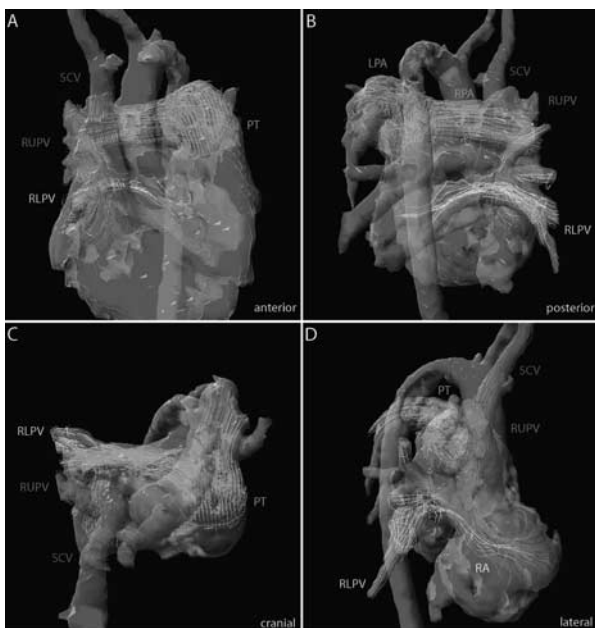
**Conclusions:** CMR can be effectively and safely performed in neonates with CHD, at time of first diagnosis even in critically ill patients. The information obtained has a major clinical impact on further management and obviates other invasive and potentially harmful examinations.

#### PW2-12

##### Selective pulmonary venous flow visualization and quantification by flow-sensitive four-dimensional cine magnetic resonance imaging facilitates and improves the accurate diagnosis of partial anomalous pulmonary venous drainage

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**Objectives:** To assess if flow-sensitive four-dimensional velocity encoded cine magnetic resonance imaging (4D VEC MRI) adds value in diagnosing patients with suspected partial anomalous pulmonary venous drainage (PAPVD).



**Methods:** In six patients with echocardiographically suspected PAPVD, anatomy was evaluated using standard magnetic

resonance imaging including angiography. Functional analysis included shunt calculations from standard flow-measurements. Furthermore, 4D VEC MRI was used for visualization of maldraining pulmonary veins and quantification of flow via the maldraining veins and interatrial communications, if present.

**Results:** In all patients, the diagnosis of PAPVD was confirmed by standard magnetic resonance imaging. Shunt volumes ranged from 1.4:1 to 4.7:1. Drainage sites were the superior caval vein ( $n=5$ ) or the vertical vein ( $n=1$ ). Multiple maldraining pulmonary veins were found in three patients. Pulmonary arteries and veins could be clearly distinguished by selective visualization using 4D VEC MRI. Flow measured individually in maldraining pulmonary veins ( $n=6$  patients) and across the interatrial communication ( $n=3$  patients) revealed a percentage of the overall shunt volume of 30 to 100% and 58 to 70%, respectively. Different flow characteristics and directions of maldraining pulmonary veins and across the interatrial communication could be visualized during ventricular systole and diastole.

**Conclusion:** Selective visualization of individual vessels and their flow characteristics by 4D VEC MRI facilitates to distinguish adjacent pulmonary arteries and veins and thus improves accurate diagnosis of maldraining pulmonary veins. By detailed quantification of shunt volumes additional information for planning of treatment strategies is provided. This method adds clinical value and might replace contrast-enhanced magnetic resonance angiography in these patients in the future.

Figure: Red streamlines represent flow of maldraining right upper pulmonary vein (RUPV) entering the right atrium (RA) together with blue streamlines of the superior caval vein (SCV) (panel A and B).

Green streamlines in the pulmonary trunk (PT), right and left pulmonary artery (RPA and LPA) allow distinguishing peripheral pulmonary arteries (green) and veins (yellow and red, panel C). Yellow streamlines of correctly draining right lower pulmonary vein (RLPV) merge with red streamlines representing a left to right shunt across the interatrial communication and thus functional maldrainage during ventricular systole (panel D).

#### PW3-1

##### Correlation of maternal flecainide levels and therapeutic effect in fetal supraventricular tachycardia

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**Background:** Transplacental flecainide is an established therapy for fetal supraventricular tachycardia (SVT). There is a paucity of data relating to the dose-response relationship of flecainide for this indication.

**Objective:** To review the relationship between flecainide levels and arrhythmia control for fetal SVT between January 1997 and December 2008 at a tertiary fetal cardiology unit.

**Methods:** Review of records of fetal SVT cases. Other arrhythmias, for example, atrial flutter, were excluded.

**Results:** Flecainide was initiated at dose of 100 mg three times a day. 27 fetuses were treated with flecainide at median gestation 31 weeks (range: 21–38) for persistent ( $n=19$ ) or intermittent SVT ( $n=8$ ). Median fetal heart rate was 250/min (range: 215–316). 11 fetuses were hydropic at initiation; 1 had long ventriculo-atrial tachycardia. Flecainide was administered first-line therapy in 17 and second-line in 10 with no significant. 19/27 (70%) fetuses converted to sinus rhythm. The median time to conversion was 3.5 days (range: 2–48). There were no fetal deaths on flecainide.

The therapeutic range for flecainide was 200–700micrograms/litre. The median flecainide level at reversion to sinus rhythm was 460 (range 250–840). 16/18 responders had a flecainide level within the therapeutic range, 2 were supra-therapeutic. The median flecainide level was 360 (range: 150–840) in fetuses who did not cardiovert and was not significantly different from responders. All non-responders achieved a therapeutic flecainide level during therapy.

Pharmacological cardioversion occurred in 10/11 (91%) hydro-pic fetuses at a median of 3.5 days (range: 2–11) and in 9/16 (56%) non-hydro-pic fetuses at a median of 4 days (range: 2–48). There was no significant difference between the flecainide levels in the hydro-pic versus non-hydro-pic groups. There was no significant difference in flecainide levels in non-hydro-pic fetuses who responded to flecainide (median 455, range 250–700) versus the non-hydro-pic responders (median 645, range 340–840).

**Conclusions:** Maternal flecainide levels do not predict cardioversion in the fetus with SVT. The clinical response to flecainide appears good, particularly in hydro-pic fetuses. Differences in fetal response may be related to placental transfer or electrophysiological properties of the arrhythmia rather than to the maternal blood flecainide level. The fetal safety profile of flecainide appears good.

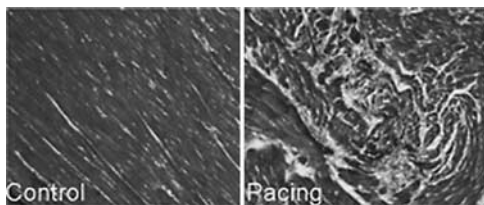
### PW3-2

#### Chronic cardiac pacing induces structural and functional remodeling in the porcine postnatal developing heart

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**Introduction:** The impact of chronic cardiac pacing in paediatric patients requires further elucidation. We investigated the effect of chronic pacing during porcine postnatal cardiac development.

**Methods:** Nineteen new-born pigs with normal LV function were divided into controls (no pacing; n = 5) atrio-right ventricular pacing (RVP, n = 7) and atrio-left ventricular pacing (n = 7). After a six fold increase body weight, we performed echocardiography, left and right ventricular myocardial biopsies. Only data from the control and RVP groups were analyzed.



**Results:** Compared to controls, RVP animals showed a significant ( $p < 0.05$ ) decrease in LV ejection fraction, and a significant increase in left atrial size. RV pacing resulted in myocardial disarray and in a significant increase in the myocardial fibrosis score (figure). A significant increase in stress-response kinase ERK1/2 was also observed ( $p < 0.05$ ). No significant difference in global or regional expression of SERCA2a ATPase, total caspase 3 and connexin 43 was identified. Finally, we found a dramatic alteration in myocardial mitochondrial function, with a significant reduction of maximal oxygen consumption and a positive response to cytochrome c test ( $p < 0.05$ ), which indicates an alteration of the outer mitochondrial membrane integrity.

**Conclusions:** RVP during postnatal cardiac development in the porcine heart results in marked changes in cardiac structure and function. The analysis of the LV pacing group may answer the question whether the negative impact of ventricular pacing can be minimized by LV pacing.

### PW3-3

#### The lumenless 4.1 Fr transvenous Pacemaker electrode (Medtronic 3830) in patients with congenital heart defects – a single center experience with 39 patients

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**Introduction:** Transvenous pacing is state-of-the-art in adult patients whereas epicardial pacing may have advantages in children or in congenital heart disease (CHD). Due to its very slim design (4.1 Fr), the recently introduced model 3830 lead (Medtronic Inc., MN, USA) is a very attractive candidate for transvenous lead delivery even in very young and CHD patients. We report our first experience with this bipolar fixed-screw, steroid-eluting lumenless pacing lead in this group using adjusted application techniques to avoid vascular access trauma.

**Methods:** All procedures were done in conscious sedation except for 2 cases with general anaesthesia. Venous access was obtained in standard fashion via the cephalic vein. For lead placement we either used the original steerable catheter (7 Fr) without an outer sheath or in patients  $< 20$  kg, a modified cut-off peel-away 5 Fr Attain Select II catheter (Medtronic) without additional sheath. Subcutaneous Generator placement was performed in a standard fashion.

	Implantation	
	Threshold	Sensing
atrial n = 35	$0.7 \pm 0.36$ V (0.3–2.1)	$3.0 \pm 1.64$ mV (1.4–10.0)
ventricle n = 23	$0.5 \pm 0.17$ V (0.25–1.0)	$12 \pm 7.4$ mV (2.8–30.0)
	Last Follow up	
	Threshold	Sensing
	$0.5 \pm 0.24$ V (0.25–1.25)	$2.8 \pm 2.12$ mV (1.4–8.3)
	$0.75 \pm 0.78$ V (0.25–3.75)	$15 \pm 10.16$ mV (4.0–31)

**Results:** 58 Electrodes were implanted in 39 patients: age was 12.8 y (2.1–70) and weight 43 kg (9.7–114), 7 patients  $< 20$  kg. Ventricular leads were successfully placed in RVOT or RV apical septum. There were no lead displacements acutely and no procedural complications. No failure to capture or sense was observed during implant. Follow-up was 392 d (8–1209). Initial pacing thresholds were low in all patients and remained stable during follow-up except in 3 leads with significant increasing values. In one patient this led to change of ventricular electrode after 955 days.

**Conclusions:** The 3830 pacing lead can be successfully implanted in the CHD population with good short- and midterm performance. We used modified delivery techniques that minimize the access size to 5–7 Fr, to tap the full potential of the very slim 4.1 Fr design even for small patients. Of course, long-term data on greater patient collectives are needed to confirm these results. The electrode has several advantages in (complex) CHD patients.

### PW3-4

#### Unravelling The Mechanisms Behind The Hemodynamic And Autonomic Remodeling Induced By Orthostatic Training

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**Introduction:** Neurocardiogenic reflex syncope (NRS) is a common clinical entity resulting from excessive reflex autonomic responses, particularly during orthostatism. Therapeutic options are controversial and of limited effectiveness. Tilt-training (TT) has been advocated as a promising tool to treat these patients (P). However, its mechanisms of action and clinical impact remain elusive.

**Aim:** to characterize hemodynamic, autonomic and baroreflex responses during a TT-program in NRS-P refractory to conventional measures.

**Methods:** 28P (50% male,  $41 \pm 14$  yrs) without structural heart-disease, with recurrent NRS documented by tilt-testing (cardioinhibitory 50%, mixed 35%, and vasodepressor 15%). The TT-program included 9 tilt-sessions (3 times/week, 30 min,  $60^\circ$  in 6 sessions and  $70^\circ$  in 3 sessions) under continuous ECG and blood pressure monitoring, combined with daily home orthostatic self-training (20 min with back support) and  $10^\circ$  head-up during sleep. Systolic-volume, cardiac-output, total-peripheral-resistance ( $\text{dyne} \cdot \text{s}/\text{cm}^5$ ), baroreflex-sensitivity and heart-rate variability were computed. P were followed during  $24 \pm 12$  months (1st month and every 6 months).

**Results:** After the TT-program there was a significant increase of total-peripheral-resistance ( $1485 \pm 225$  vs.  $1591 \pm 187$ ,  $p < 0.05$ ), with a reduction of its standard deviation ( $206 \pm 60$  vs.  $150 \pm 42$ ,  $p < 0.05$ ). Variability studies using auto-regression analysis showed an increased overall autonomic activity, reflected by a progressive increase in the variability indices (LF  $544.08 \pm 146$  vs.  $914.64 \pm 225$ ,  $p < 0.05$ ; HF  $5.32 \pm 0.7$  vs.  $7.42 \pm 0.8$ ,  $p < 0.05$ ). This increase can also be seen through changes in the baroreflex effectiveness index ( $61.06 \pm 20$  vs.  $69.70 \pm 17$ ,  $p = 0.08$ ). Recurrence of syncope occurred in 5P (19%), with a significant reduction in the number of episodes ( $4.0 \pm 3.2/P$  in the 12 months before TT vs.  $1.4 \pm 0.8/P$  post-TT,  $p < 0.05$ ).

**Conclusions:** In refractory NRS, TT may be an effective option, with long-term benefits due to a better orthostatic tolerance through three mechanisms: increased vasoconstrictor reserve and its lower variability combined with increased overall autonomic tone and baroreflex activity.

### PW3-5

#### **Pregnancy and neonatal complications in women with congenital cardiac disease – a long-term follow-up study**

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**Background:** Pregnancy in women with congenital cardiac disease (CCD) become more frequently due to the progress in the field of diagnostic techniques and surgical interventions. Data regarding the pregnancy complications and neonatal outcome are limited.

**Methods and Results:** 267 pregnant women with partly complex CCD were analysed in one of the largest single centre cohorts in Germany. The median age was 27 years. The frequency of maternal and neonatal outcome and complications were monitored.

The main cardiac complications were arrhythmias (12%) and heart failure (10%). 29% of these symptomatic arrhythmias were treated. 44% of the patients with complex heart diseases, but also 23% with simple lesions lost at least one functional class. 2% had thromboembolic events. Two mothers died within one year after delivery.

The most prevalent neonatal complications were premature birth (12%) and small for gestational age (8%). According to the Federal statistical office of Germany the rate of prematurity was twice as high as in the normal population. 24% of the premature babies were seen in the patients with complex cardiac diseases. Congenital cardiac defects in the neonates were seen in 5% of all pregnancies.

Additionally most women were contacted by mail and asked to fill in the health situation and medical care in/after pregnancy for a long term follow-up.

62% of the women answered the questionnaire. Fortunately most late survivors were active and had a good quality of life. With regard to the functional class of Perloff 58% of the patients considered themselves as healthy and 30% had a reduced functional class after pregnancy. 87% were satisfied with the medical surveillance.

**Conclusion:** Successful pregnancy can be achieved in most women even with congenital cardiac disease. The rate of cardiovascular morbidity and premature delivery is increased especially in patients with complex heart diseases but also in patients with simple lesions. As a result patients should be monitored closely by a multi-disciplinary health care team that includes obstetricians, cardiologists, pediatric cardiologists and obstetric anaesthesiologists.

### PW3-6

#### **Impaired whole blood coagulation and platelet function in patients with cyanotic congenital heart disease**

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**Objectives:** Many patients with cyanotic congenital heart disease (CCHD) suffer from hemostatic and coagulation abnormalities resulting in both – bleeding diathesis and thromboembolic events. Bleeding diathesis may be related not only to coagulation abnormalities but also to reduced platelet count and function. Measurements of the velocity and firmness of clotting by thromboelastometry have sporadically been described as a useful method for detection of hypercoagulability. This study was performed to assess the impact on thromboelastography and platelet aggregometry in patients with CCHD.

**Methods:** 55 consecutive patients with CCHD presenting in our center were studied. Blood samples were taken for detailed laboratory analysis including modified thrombelastography. Blood cells were counted on a Sysmex XE2100 analyser and the Multiplate™ system was used for measuring the whole blood impedance platelet aggregometry. Thrombelastometry was performed using tissue-factor as activator (EXTEM) and tissue-factor plus platelet inhibitor cytochalasin (FIBTEM) with the ROTEM™ system. Concomitant medication with potential impact of blood coagulation and platelet function were noted.

**Results:** Median hematocrit (Hct) was 57% (range: 42–78%). There are negative correlations between Hct-levels and platelet counts ( $r = -0.5372$ ), platelet aggregation after activation with ADP ( $r = -0.4103$ ), arachidonic acid ( $r = -0.5249$ ) and TRAP ( $r = -0.3327$ ) similarly to EXTEM-analysis (maximum clot firmness -MCF-  $r = -0.5729$ ) and the alpha angle ( $r = -0.6978$ ). The median MCF-value in the FIBTEM test was 9 mm, showing fibrinogen components of blood coagulation after in-vitro inhibition of platelet function. Hence, 50% of all investigated patients with CCHD had fibrinogen equivalents below the reference range and in patients with higher Hct-levels FIBTEM-values were even lower (see table).

	<b>Median [25.–75. percentile] (Hct &lt; 57%)</b>	<b>Median [25.–75. percentile] (Hct ≥ 57%)</b>
Platelet count [1/nl]	178 [80–313]	120 [56–297]
ROTEM EXTEM MCF [mm]	57 [43–68]	50.5 [31–68]
ROTEM EXTEM alpha angle [°]	66 [51–75]	52 [32–77]
ROTEM FIBTEM MCF [mm]	10.5 [3–23]	7 [2–20]
Multiplate ADP-ind. Aggregation [AU*min]	492 [56–879]	320 [200–844]
	<b>Kruskal-Wallis test significance level p</b>	<b>Reference range</b>
	p = 0.0013	200–400
	p = 0.0025	50–72
	p = 0.0001	63–83
	p = 0.0534	9–25
	p = 0.0829	534–1220

**Conclusions:** Complex hemostatic abnormalities, including thrombocytopenia and suppressed platelet function, are common in CCHD. Thrombelastometry and platelet aggregometry are useful instruments for assessment of the whole blood coagulation and platelet function also in these patients. Thrombometry demonstrated a tendency for inferior clot formation dynamics (alpha angle in EXTEM), decreased fibrinogen and disturbed clot polymerization (small MCF in FIBTEM), even though no tendency for hypercoagulability could be found. Particularly with regard to decisions about anticoagulation or antiplatelet therapy, in patients with CCHD this analytical methods could become relevant.

### PW3-7

#### Long-term neo-aortic growth in arterial switch operation versus Ross procedure. Is there a difference?

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**Introduction:** Both arterial switch operation (ASO) in D-transposition of the great arteries and Ross procedure (ROSS) in aortic valve disease include previously pulmonary tissue in high-pressure systemic circulation. This may lead to proximal aorta dilatation and malfunction. The aim of the study was to compare long-term neo-aortic growth/function in these patients.

**Patients and methods:** Compared were patients with ASO vs ROSS: 85 vs 48 patients, sex: 56M/29F vs 31M/17F, median age at surgery: 10 days vs 12.9 years ( $p < 0.0001$ ), long-term follow-up (FU): median 11.6 vs 8.2 years. In patients repeated longitudinal echocardiographic proximal neo-aortic diameters (1.annulus (ANN), 2.sinuses (SIN), 3.sino-tubular junction (STJ)) were measured and converted to Z-scores according to normal values (measured in 702 healthy controls). Statistical analysis of ASO and ROSS was performed at 5, 10 and >10 years after surgery.

**Results:** Significant difference in ASO vs ROSS median Z-score was found: 1.ANN (1.8 vs 2.7 –  $p < 0.001$ ), 2.SIN (2.2 vs 3.0 –  $p = 0.0022$ ), 3.STJ (0.2 vs 3.9 –  $p < 0.001$ ). Comparing ASO vs ROSS during FU: at 5 and 10 years after surgery showed significant dilatation of: 1.ANN (1.5 vs 2.9 –  $p < 0.001$ ) and 1.5 vs 2.5 –  $p = 0.0021$ ), 2.SIN (2.0 vs 3.5 –  $p = 0.01$  and 2.0 vs 2.9 –  $p = 0.01$ ), 3.STJ (–0.05 vs 4.0 –  $p < 0.001$  and –0.35 vs 3.9 –  $p < 0.0001$ ). At >10 years after surgery there was no statistical difference in: 1.ANN (2.7 vs 2.5 –  $p = 0.68$ ), 2.SIN (2.8 vs 2.9 –  $p = 0.5$ ), only in 3.STJ (1.2 vs 3.9 –  $p < 0.0001$ ). Neo-aortic regurgitation was present in 41.8% ASO vs 64% ROSS ( $p = 0.0013$ ). In ROSS compared to ASO was regurgitation more often present with dilated (Z-score >2) 1.ANN ( $p = 0.002$ ), 2.SIN ( $p = 0.003$ ), but not with dilated 3.STJ ( $p = 0.34$ ).

**Conclusions:** Proximal neo-aortic dilatation is significantly greater in patients after ROSS compared to ASO. Measured diameters show no progression during FU in patients with ROSS, also in ASO during first 10 years of FU; though >10 years after surgery there is a significant progression in ASO patients. This may be associated with the different age at surgery in both groups (new-born in ASO vs >10 years in ROSS).

### PW3-8

#### Long term follow up of adolescent and adult grown up congenital heart disease (GUCH) patients after modified Fontan operation

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**Introduction:** Since the introduction of single ventricle palliation more patients reach adulthood and need specialized supervision and therapy. We analyzed hemodynamics, physical performance and morbidity in GUCH patients after modified Fontan operation.

**Methods:** Seventy patients, who underwent Fontan operation between 1991 and 2010, reached adolescent (15–18 years) or adult age (range: 15–50, median 27 years) during the median follow up of 10.4 (range: 0.6–19) years; 20 of them were older than 30 years. Seventeen patients were operated on as adults (16–37 years). The intraatrial modification was performed in 38 patients and extracardiac Fontan operation (ECFO) in 32. The hemodynamics was analyzed by heart catheterization and MRI. The cardiopulmonary capacity was tested by spiroergometry with monitoring of the oxygen consumption capacity ( $VO_{2max}$ ). Necessity for cardiac medication and the incidence of arrhythmias were checked.

**Results:** There were three late deaths (mortality 4.3%). Patients who underwent heart catheterization ( $n = 43$ ) showed low pulmonary artery pressure (median 11 mmHg) and low transpulmonary gradient (median 6 mmHg). The cardiac index, measured by MRI ( $n = 29$ ) was in median in adolescents better than in adults (3.1 vs. 2.4 l/min/m<sup>2</sup>,  $p = 0.027$ ).  $VO_{2max}$  decreased significantly during the follow up ( $27.1 \pm 7.7$  ml/min/kg (59.9 ± 17.0%) early vs.  $20.2 \pm 7.8$  ml/min/kg (54.7 ± 17.9%) late,  $p = 0.005$ ). Thirteen patients (19%) developed tachyarrhythmias; 11 patients, all with intraatrial Fontan operation, required a permanent pacemaker due to bradyarrhythmias. Medical treatment of heart failure with more than two drugs was necessary in 41 patients (59%). All patients were on an anticoagulation regime. No clinically relevant thromboses were noted but one thromboembolic event occurred in one patient after intracardiac Fontan. No transplantations were necessary in adult patients after Fontan operation during current follow up.

**Conclusions:** Grown up patients after lateral tunnel or extracardiac Fontan operation reach adulthood with stable hemodynamics and low morbidity. The incidence of arrhythmias after ECFO is lower. Regular checks of the patients' physical exercise capacity and hemodynamics are necessary to optimize the cardiac medication for progressive heart failure and to identify candidates for later heart transplantation.

### PW3-9

#### Intellectual outcome in children after complex cardiac surgery: Arterial switch operation vs. total cavopulmonary connection

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**Introduction:** For both groups, patients after arterial switch operation (ASO) for d-transposition of the great arteries (d-TGA) and children with functional single ventricle after total cavopulmonary connection (TCPC), a lower intellectual function compared to the general population has been reported (e.g. ASO: Bellinger et al., 2004, Karl et al., 1999; TCPC: Forbess et al., 2001, Sarajuuri et al., 2007). The purpose of the present study was to compare intellectual outcome of these two groups of patients.

**Methods:** In 29 patients after ASO for d-TGA (aged 4 to 12 years; mean age  $7.5 \pm 2.5$  years) and 89 patients with functional single ventricle after TCPC (aged 3 to 12 years; mean age  $7.5 \pm 2.5$  years) intelligence was assessed by the Kaufman-Assessment Battery for Children (K-ABC; German version). The K-ABC measures two aspects of intellectual function, that is fluid intelligence ("mental processing scale" or "intelligence scale") and crystallized intelligence ("achievement scale") on different scales. For analysis, the mean test scores of the groups were compared to each other.

**Results:** Overall, patients after TCPC achieved lower mean scores on all subscales of the K-ABC than children after ASO: On the fluid intelligence scale, they scored descriptively but not significantly lower ( $93.2 \pm 12.2$ ) than patients after ASO ( $96.1 \pm 10.1$ ,  $p = .13$ ). Aside from two subtests – "gestalt closure" and "number recall" – they scored descriptively lower on all subtests of this scale. On the crystallized intelligence scale, they achieved a mean score of  $92.1 (\pm 14.8)$ , which was significantly lower than the mean score of children after ASO ( $97.3 \pm 11.6$ ,  $p < .05$ ). They showed a descriptively worse performance on all subtests of this scale.

**Conclusions:** Patients with functional single ventricle after TCPC achieve lower mean fluid and crystallized intelligence scores than children after ASO for d-TGA. These findings are consistent with the report of Brosig et al. (2007). Patients after TCPC especially require careful observation of neurodevelopmental outcome and appropriate interventions when indicated.

### PW3-10

#### Induction therapy with Daclizumab in pediatric heart transplantation

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Daclizumab is a humanized monoclonal antibody which binds with high affinity to the Tac subunit of the IL-2 receptor complex. Effective immunosuppression with Daclizumab in adult patients encouraged the initiation of the administration of Daclizumab as induction therapy in pediatric heart transplantation.

Sixteen patients (9 boys, 7 girls, age 8.7 yrs, BMI  $1.75 \pm 0.52$  m<sup>2</sup>), received Daclizumab as induction therapy in a dosis of 1 mg/kg intravenously perioperatively and on day 7 and 21 after orthotopic heart transplantation. Additional immunosuppression was cyclosporine (CsA,  $n = 14$ ) or tacrolimus (TAC,  $n = 2$ ), mycophenolate mofetil (MMF) and prednisolone. Prednisolone was tapered rapidly in the first six months after heart transplantation. The administration of Daclizumab was not associated with any side effect. Owing to the blockade of the IL-2-receptor the dosage of calcineurin inhibitors could be reduced leading to less renal and hepatic toxicity. Instead of aiming at CsA trough levels of 350–400 ng/ml/TAC trough levels of 12–15 ng/ml in the first weeks after transplantation we reduced to 250 in the CsA group and to 10 in the TAC group. CD25+ T-lymphocytes began to be re-expressed after 2–3 months after administration of Daclizumab.

In a mean follow-up time of 50.2 months no acute or chronic episode of rejection could be experienced. The incidence of opportunistic infections was not elevated (5 bacterial, 4 viral and 3 fungal infections which responded well to adequate treatment). No de novo malignancies, especially no lymphoproliferative disease (PTLD) was noticed. Actually patient and graft survival is 100%.

Our results show that immunoprophylaxis with Daclizumab induction therapy in pediatric heart transplantation is safe, effective and well tolerated and does not lead to increased opportunistic infections or malignancies. The reduction of calcineurin inhibitors led to less calcineurin related side effects and raised the quality of life of transplanted patients.

### PW3-11

#### Results of pediatric heart transplantation after 15 and 20 years: How are they now?

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**Introduction:** Pediatric heart transplantation (HTX) has been a well respected procedure for end-stage heart disease for the past 30 years. Besides the improved survival in the last years, the long-term outcome is influenced by rejections, posttransplant lymphoproliferative disease (PTLD), chronic renal insufficiency, graft coronary artery disease and retransplantation. Therefore, we focused on morbidities in survivors of more than 15 years after HTX.

**Methods:** Patients (pts) with a pediatric heart transplantation before 1995, in our institution, were retrospectively reviewed.

**Results:** There were 30 pts transplanted more than 15 years ago, of those, 6 were more than 20 years ago. The mean age at time of HTX was 1.83 years (range 0.08–15.6). 23 pts (80%) were under the age of two years. Diagnosis which lead to HTX was: a complex congenital heart defect (66.6%) and cardiomyopathy (23.4%). There were 48 rejection episodes, most of them in the first 6 months after HTX. 3 pts suffered from PTLD, 2 had a relapse. All pts have a mild chronic renal insufficiency with a mean GFR of  $92.4 \text{ ml/min/m}^2 (+/-19.78)$  after 15 and  $82.06 \text{ ml/min/m}^2 (+/-28.48)$  after 20 years. One pt needed kidney transplantation. 2 pts had severe coronary artery disease, followed by a re-HTX in one pt after 20 years.

13 pts died. There was an early death within one month after transplantation in 66.6%, mostly due to graft failure ( $n = 5$ ). Late death occurred in 33.4%, after rejection in 3 pts and PTLD in 1 pt. Of the 30 pts, 58.7% are still alive after 20 years. Immunosuppression is based on a calcineurin-inhibitor in 52.9%, with a majority of 77.7% with tacrolimus. The remaining pts (47.1%) are treated with an CNI-free mTOR-inhibitor regime. All pts have a steroid-free regime at 3–6 months after HTX.



**Conclusion:** Morbidities in pediatric heart transplantation are challenging: Rejections mostly occur in the first 6 months after HTX. Chronic renal insufficiency, malignancy and coronary artery disease will add on by time. Tailored use of immunosuppressants, as influencing factor, might be the key factor in reducing the morbidities.

### PW3-12

#### **Spanish Collaborative Group in Pediatric Ventricular Assisted Device: Experience with the Berlin Heart Excor**

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**Introduction:** Ventricular Assisted Device (VAD) is an option in severe heart failure in pediatric patients as bridge to recovery or heart transplantation. Lengthy waiting lists are an issue in young patients, and long term devices need to be specifically evaluated in children, focused on avoiding complications, improving transplant survival or recovery without sequelae. Here we review our national experience with the Berlin Heart (BH) Excor Pediatric Device.

**Methods:** From 2006 to January 2011 all patients and all institutions with implants, 5 public Spanish children's hospitals, enrolled in a collaborative group.

**Results:** 22 patients from 1 month to 15 years age (mean 3,1 year), 9 (41%) infants, with a weight from 3,5 to 60 kg (mean 14,8 kg), with 12 (55%) less than 10 kg were included. 16 (73%) were cardiomyopathies (4 myocarditis) and 6 (27%) postoperative congenital heart disease (4 univentricular with 2Fontan, 1 Shone Syndrome and 1 ALCAPA). ECMO was previously used in 11 (50%) with a mean duration of 10 days (range 2 to 17). 1 patient was assisted with Levitronix VAD previously. Left VAD was the initial assistance in 13 (59%) and in 2 of them switch to BiVAD. Atrial cannulation was required in 2 cases (9%) and transitory interposition of an oxygenator in one. Mean duration of assistance was 61 days (median of 36 days and range of 7 days to 7 months). The complications found were: severe hemorrhagic (50%), thromboembolic (18%), infectious (36%), neurologic (45%), thrombus and replacement device (18%). 13 patients (59%) were transplanted, 8 (36%) death. Higher death rate were found in structural disease (3/6 = 50%) and previous ECMO (5/12 = 63%). One patient continues with the VAD. Explantation due to recovery was not achieved. Finally 12 patients were discharged from ICU and 11 (50% from total and 84% post transplant) survived.

**Conclusion:** In our collaborative experience BH is a VAD that provides medium-to long-term support in infants and children as bridge to heart transplant, with posterior high survival. Complications and death during the assistance remain an issue, especially in structural heart disease and previous ECMO patients.

### PW4-1

#### **Fetal Congenital Heart Disease in Twins with fully identified Zygosity: genetic or epigenetic? That is the question perhaps answered by an international multicenter study of 144 monozygotic twins**

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Twins allow understanding of malformations and provide data of their cause and genetic component. The incidence of cardiac malformations (FHD) is higher in monozygotic (MZ) than in dizygotic and singletons. MZ have a similar environment. This multicenter prenatal study is the first to collect a large number of MZ, 144 sets of MZ with fully identified zygosity. Acardiac and FHD in twin to twin transfusion were excluded. Our purposes were to answer the following questions: 1-what was rate of concordance, genetic or not? 2-what were the phenotypes of the FHD? (we used Clark's classification) 3-could we suspect any influence of epigenetic factor, in particular placentation? Prenatal diagnosis of the FHD was performed using Dopplerechocardiography (2D). Extremes for gestational age were 16–35 weeks. Whenever intra-uterine death or termination of pregnancy occurred, fetopathological examination was done. All neonates had 2D. Fetal karyotypes were obtained with microdeletion 22q11 testing. Zygosity was assessed taking account fetal gender, gonosomal complement, placental phenotype and microsatellite analysis. Placental examination was performed. Only 16 sets of MZ presented each a FHD, but there were discordant, however there was one exception, one MZ presented twins with aortic and mitral atresia. We also found that an extracardiac malformation (ECM) was found in the twin with FHD in 18% of the MZ. The answer to first question is discordance for FHD and the ECM when present. The FHD phenotype showed that type II (flow lesions) and type I (conotruncal malformations) accounted for 75% of the phenotype with a predominance of the abnormal flow, followed by type IV (atrioventricular canal) and only two were type VI (looping anomalies). The placentation showed relevant information, there was an abnormal incidence of velamentous or marginal cords. This result has been already reported in MZ with ECM (Machin, G.A., 1996). Velamentous or marginal cord were found significantly more frequent in MZ with FHD. In conclusion this multicenter study in MZ showed discordance in FHD. This observed discordance probably occurs as a postzygotic event. We have strong indication that one of the epigenetic factors maybe abnormal cord insertion, others are searched such as factors of methylation.

## PW4-2

### Heart valve tissue-engineered matrices attenuate monocyte binding and procoagulant responses in human endothelial cell cultures exposed to *S. aureus*, *S. sanguis* and *S. epidermidis*

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**Introduction:** Infective endocarditis (IE) remains a serious complication after heart valve replacement. Autologous valves constructed by matrix-based tissue engineering are under investigation to increase biocompatibility. The impact of the underlying matrix on the risk to develop IE is not known. IE is characterized by bacterial adhesion and subsequent interactions of disseminating bacteria with endothelial cells (ECs) and monocytes, evoking endothelial proinflammatory and procoagulant activity, leading to heart valve destruction.

**Methods:** In the present study we therefore have seeded human ECs on a fibrin vs. collagen gel matrix, and, at confluence, infected them with *Staphylococcus aureus*, *Streptococcus sanguis* and *Staphylococcus epidermidis*.

**Results:** Especially *S. aureus* infected ECs grown on fibrin (4.2% of the inoculum) and collagen (3.7%) matrices, even more than on ECs grown on non-coated plates (1.2%;  $p < 0.01$ ). This was associated with higher monocyte adhesion (61% on fibrin and 43% on collagen) than in the control cultures (30%,  $p < 0.01$ ), even when the EC surface expression of ICAM-1 and VCAM-1 remained comparable. The collagen matrix attenuated the *S. aureus* induced MCP-1 expression 2.0 fold, compared to the non-coated control ECs. This reduction prominently coincided with a 4.2–5.0 fold reduction in the procoagulant activity, triggered in ECs grown on non-coated wells, as a consequence of tissue factor expression by ECs, further stimulated by EC-bound monocytes. Moderate responses were seen upon infection with *S. sanguis* and *S. epidermidis* for both gel matrices.

**Conclusions:** Thus, even when fibrin and collagen gel matrices equally increase bacterial adhesion, and subsequent monocyte adhesion to infected ECs, these matrices modulate EC responses to these stimuli, resulting in attenuated cytokine production and attenuated adherent monocyte-dependent tissue factor production by the ECs. Further investigations will need to confirm that also in vivo, EC-matrix interactions can attenuate EC responses to bacteria and inflammatory cells to reduce IE at infected endovascular sites.

## PW4-3

### Inhibition of calcineurin-signalling attenuates RV adaptation to pressure load in mice

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**Introduction:** Right ventricular (RV) failure is an important determinant of outcome in congenital heart disease and pulmonary hypertension. RV adaptation to pressure load is characterized by calcineurin-activation and pathological hypertrophy. We tested the effects of genetic blockade of calcineurin on RV adaptation to pressure load in mice.

**Methods:** Mice with cardiac-specific inducible upregulation of Modulatory Calcineurin-Interacting Protein (carMCIPtg), the inhibitor of calcineurin-activation, were injected with tamoxifen to induce the upregulation. Next, they were subjected to pulmonary artery banding (PAB) via a left lateral thoracotomy or sham-operated (Sham) and to cardiac MRI 4 weeks after surgery. Results were compared with the response of wild type mice (WT) to PAB.

**Results:** In carMCIPtg, activation of calcineurin in response to pressure load (PAB) was successfully blocked as shown by the lack of endogenous MCIP1 upregulation (all data in Table). Also, beta-MHC upregulation in response to PAB was blocked, but not alpha-MHC downregulation. CarMCIPtg had reduced RV hypertrophy in response to PAB as shown by lesser RV weight/body weight ratio. Moreover, RV dilatation in response to the pressure load, seen in the WT mice, was attenuated. However, the sham-operated  $\alpha$ -MHC-MCIP mice also had RV dilatation, suggesting that calcineurin-signalling is required for normal RV homeostasis.

**Conclusions:** These results show that blocking calcineurin in the pressure loaded RV attenuates RV hypertrophy and dilatation, but induces RV dilatation in the unstressed RV. Hence, there is a delicate balance in calcineurin-signalling in the normal versus the pressure loaded RV. Intervention in this balance may be selectively beneficial in stressed RV's.

	carMCIPtg		Wild Type	
	Sham	PAB	Sham	PAB
Gene Expression (relative to reference gene)				
Endogenous MCIP1	1.0 $\pm$ 0.3	0.9 $\pm$ 0.3†	1.0 $\pm$ 0.1	4.6 $\pm$ 1.3*
Beta-MHC	1.0 $\pm$ 0.3	1.3 $\pm$ 0.6†	1.0 $\pm$ 0.1	14.1 $\pm$ 4.1*
Alpha-MHC	1.0 $\pm$ 0.3	0.4 $\pm$ 0.1*	1.0 $\pm$ 0.2	0.4 $\pm$ 0.1*
RV hypertrophy (RV weight/ body weight mg/g)				
	0.8 $\pm$ 0.1	1.2 $\pm$ 0.1*†	0.9 $\pm$ 0.2	1.6 $\pm$ 0.1*
MRI derived RV volumes				
End-diastolic ( $\mu$ l)	65 $\pm$ 9	58 $\pm$ 5†	30 $\pm$ 5	39 $\pm$ 6*
End-systolic ( $\mu$ l)	32 $\pm$ 6	31 $\pm$ 4†	10 $\pm$ 2	15 $\pm$ 3*
Ejection Fraction (%)	52 $\pm$ 3	47 $\pm$ 3†	69 $\pm$ 4	62 $\pm$ 6
Cardiac output ( $\mu$ l/min/g)	333 $\pm$ 41	351 $\pm$ 28	362 $\pm$ 69	355 $\pm$ 25

N = 4–6 per group, \* $p < 0.05$  vs. Sham, † $p < 0.05$  vs. WT

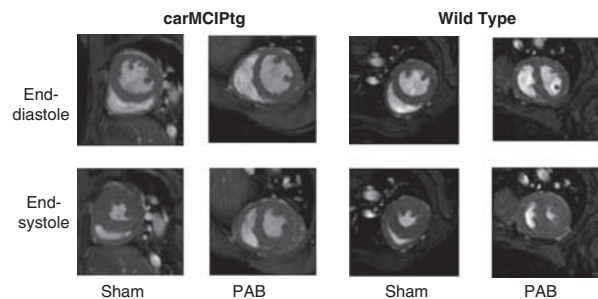


Figure: typical examples of cine MRI slices

## PW4-4

### Long term changes in retinal vasculature from Kawasaki Syndrome and its potential role in coronary risk stratification

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**Introduction:** We hypothesize that Kawasaki Syndrome (KS), a multisystem vasculitis of childhood causes longstanding changes in the retinal vasculature as a result of its attendant inflammatory effects. This is similar to the well documented changes to the retinal vasculature of adults who have traditional coronary risk factors like diabetes mellitus and hypertension.

**Methods:** We compared the retinal vascular dimensions of children in the post acute phase of KS to well matched normal controls. All subjects underwent high resolution digital retinal photography during which the diameters of all arterioles and venules coursing through a specified area one-half to one disc diameter from the optic disc were measured with a computer program (IVAN), according to a published standardized protocol. Central retinal arteriolar equivalent (CRAE), central retinal venular equivalent (CRVE), arteriole-to-venule ratio (AVR) were calculated for each retina photo.

**Results:** 18 patients with KS and 54 controls were examined. KS subjects had a mean CRAE of 157.1  $\mu\text{m}$ , a mean CRVE of 209.7 and AVR of 0.70, which were statistically different from controls who had mean CRAE of 149.3  $\mu\text{m}$  ( $p = 0.046$ ), a mean CRVE of 221.7 ( $p = 0.007$ ) and AVR of 0.66 ( $p = 0.011$ ). Cases were matched for age, gender, ethnicity, and body surface area.

**Conclusions:** KS results in substantial changes to retinal vasculature leading to a significantly wider retinal arteriolar caliber, narrower retinal venular caliber and an increased AV R ratio. Retrospective study may correlate with severity of initial inflammation and longitudinal follow up may allow for coronary risk stratification.

#### PW4-5

##### **Quality of life in asymptomatic children and adolescents before and after diagnosis of hypertrophic cardiomyopathy through family screening**

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**Introduction:** Family screening for asymptomatic hypertrophic cardiomyopathy (HCM) is motivated since the risk for sudden death can be decreased by lifestyle modifications and pharmacological treatment. Before embarking on a programme for such screening it is important to evaluate if the knowledge of having a chronic life threatening disease while being asymptomatic affects quality of life (QoL). The aim of this study was to measure QoL in asymptomatic children with HCM before and after diagnosis compared to healthy controls.

**Methods:** Quality of life was measured according to Lindström (Soz Präventivmed 1992;37:301) taking into consideration the three spheres of external, interpersonal and personal aspects. Both objective and subjective aspects were covered in all dimensions. QoL was measured using a questionnaire, before and 2 years after diagnosis. The healthy siblings of the patients served as one comparison group. In order to avoid bias from living in the same family a second comparison group of healthy children unrelated to the patients were chosen. The study group consisted of 13 children with HCM, median age 11 years (range 5–18). There were 10 healthy siblings, median age 10.5 years (range 4–16) and the other control group consisted of 31 children with a median age of 11 years (range 2–19).

**Results:** The total QoL score was similar between groups at baseline and follow-up. The total score for the personal sphere was lower at follow up in the study group compared to both control groups ( $p < 0.05$ ). The total scores for the other spheres were similar between groups at baseline and follow-up. In the interpersonal sphere the satisfaction with social support was lower in children with HCM ( $p = 0.01$ ) than in controls at baseline but not at follow up. Children with HCM had significantly more psychosomatic symptoms compared to both control groups at follow-up ( $p = 0.01$ ) but not at baseline. The subjective QoL was significantly lower in children with HCM compared to controls ( $p = 0.002$ ) at follow-up.

**Conclusion:** Family screening for HCM does not seem to negatively influence total QoL. However subjective QoL variables was lower in the study group and psychosomatic symptoms increased compared to the two control groups.

#### PW4-6

##### **Echocardiographic and Biochemical Findings of type 1 diabetic children and adolescents: Cardiovascular risk in diabetic children**

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**Introduction:** The aim of this study is to investigate the cardiovascular risk factors, to determine the atherosclerosis indicators (biochemical and radiological) and the relationship between these factors and metabolic control in children with Type 1 Diabetes Mellitus (DM).

**Methods:** The study group consists of 55 patients (31 F/ 24M) with Type 1 DM, and we selected 30 (14F/ 16M) healthy children as a control group. Level of HbA1c, lipid profile, HsCRP and adipoectin were measured. For cardiovascular examination, standard echocardiographic values (2D, Doppler), Myocardial performance index (MPI), Carotid intima media thickness (CIMT) and Flow mediated dilation (FMD) were evaluated.

**Results:** The mean age of DM patients was  $17,6 \pm 4$  years and  $16,43 \pm 4,1$  years in the control group. HsCRP values statistically significantly higher in DM patients group ( $0,21 \pm 0,31$  vs.  $0,10 \pm 0,16$  mg/l,  $p = 0,00$ ) but no difference in the levels of adinopectin ( $15,2 \pm 6,1$  vs.  $15,57 \pm 6,49$  mcg/ml) between two groups. The duration of DM has positive correlation with CIMT and negative correlation with FMD measurement. DM group has MPI abnormalities and diastolic dysfunctions of both ventricles ( $p = 0,01$ ) in Doppler measurements. FMD measurements were statistically significantly lower in DM patients group but CIMT measurements were statistically significantly higher in DM patients ( $p = 0,001$ ). FMD has negative correlation with hsCRP ( $r = -0,28$ ,  $p = 0,03$ ) and adinopectin ( $r = -0,27$ ,  $p = 0,04$ ).

**Conclusions:** Abnormal measurements in hsCRP, FMD, CIMT, diastolic and systolic dysfunction in this group show that the children with DM have cardiovascular risk in these ages. DM patients have no abnormalities in the standard echocardiographic measurements. However the advanced measurements as hsCRP, MPI, FMD and CIMT show that DM patients have cardiovascular abnormalities. As a conclusion this study shows there is an important necessity of advanced measurements techniques in the follow of the DM patients.

#### PW4-7

##### **First 20 years of paediatric heart transplantation in Sweden: Comprehensive outcome after listing for transplantation and post-transplant results 1989–2009**

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**Objective:** To evaluate, on a national basis, the outcome, including functional class, in the first generation of children with end-stage heart disease due to cardiomyopathy (CMP) or congenital heart disease (CHD) to which heart transplantation was presented as an option.

**Methods and Results:** Records of all 135 Swedish children <18 years (median age 6.5 years, range 1 day–17.6 years) listed for heart transplantation from January 1989 to December 2009 were examined. There were 74 (54.8%) CMP and 61 (45.2%) CHD patients; 34 (25.2%) were <1 year. Since 11 patients improved and were de-listed, follow up data was based on 124 patients. Waiting list mortality was 30.6% (44.4% in infants). Median waiting time until transplant in 82 surviving patients was 57 days (0–585 days). At median post-transplant follow up of 5.9 years (0.03–20.1 years), mortality was 18/82 (22%) and actuarial survival was 92.4% at one year, 82.1% at 5 years, 76.2% at 10 years, and 57.9% at 15 years. Survival after listing was 63.9% at one year, 58.5% at 5 years, 52.3% at 10 years and 39.5% at 15 years. Post-transplant complications included rejections in 34.1%, malignancies in 12.2%, renal failure in 8.5%, coronary artery vasculopathy in 6.1% and re-transplantation in 4 patients (4.9%). Among 64 survivors 84.3% had an excellent or good functional class whereas, in 15.7%, there were important complications including poor prognosis.

**Conclusion:** A high waiting-list mortality and some post-transplant attrition precluded >50% of this population from reaching adulthood. Functional class in survivors is generally good.

#### PW4-8

##### Cardiac anomalies in Noonan syndrome: experience in 64 patients

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**Introduction:** Noonan Syndrome (NS) is a variagate autosomal dominant disorder, related to mutations in the genes of RAS/MAPK pathway. NS is frequently associated with short stature, facial dysmorphism, congenital heart defects (CHD) (70–80% of cases), impaired blood clotting, lymphatic dysplasia, learning problems. Common CHD are pulmonary valve stenosis (PVS), hypertrophic cardiomyopathy (HCM) and septal defects.

**Patients and methods:** We describe cardiac involvement and genotype in a cohort of 64 patients born from 1986 to 2010, who had a clinical diagnosis of NS in our centre.

Genotype	N (%)	CHD (n,%)	PVS (n)	HCM (n)	Septal defects (n)	Other (n)
PTPN11	32 (50)	28 (44)	22	5	6	1
SOS1	8 (12.5)	6 (9)	5	0	1	0
KRAS	0	0	0	0	0	0
RAF1	3 (4.7)	3 (4.7)	1	2	0	0
BRAF	1 (1.5)	1 (1.5)	0	0	1	0
SHOC2	2 (3.1)	2 (3)	1	1	0	1
Unknown/under investigation	18 (28)	15 (25)	8	2	1	1
Sum	64 (100)	55 (86)	37	10	7	2

**Results:** We evaluated 64 patients with clinical diagnosis of NS; in 46 the genotype is known. Fifty-five patients (86%) have

cardiac anomalies, in 41 of these the molecular diagnosis was made as shown in the table. Fifteen (27%) patients needed surgical or interventional treatment, which was successful in 14 (93%). We also identified a subgroup of 17 (31%) patients in whom NS was suggested by the presence of minor cardiac anomalies.

**Conclusion:** In our series the presence of CHD in NS is higher than that previously reported and is frequently associated with PTPN11 genotype. Unlike what is generally thought we observed that cardiac surgery or interventional treatment are highly effective. One third of this population has insignificant cardiac involvement, which however lead to the genetic diagnosis.

#### PW4-9

##### Central and Peripheral Systolic Blood Pressure in Healthy Children and Adolescents and in Patients with Juvenile Hypertension

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**Basics:** The peripheral systolic blood pressure (SBP<sub>brach</sub>) is normally higher than the central systolic blood pressure (SBP<sub>ao</sub>) due to the pulse pressure amplification. The arterial stiffness increases with the normal aging of the aorta. Its consequence is the early wave reflection and increased SBP<sub>ao</sub>. Recent studies proved that SBP<sub>ao</sub> more strongly relates to cardiovascular events than SBP<sub>brach</sub> does in adult patients. We missed data about physiological changes of SBP<sub>ao</sub> in healthy children and adolescents.

**Aims:** 1. To assess the physiological changes of SBP<sub>ao</sub> and SBP<sub>brach</sub> in healthy children and adolescents. 2. To determine the relationship between SBP<sub>ao</sub> and SBP<sub>brach</sub> in patients with juvenile hypertension (JH) and compare it with control groups.

**Methods:** SBP<sub>ao</sub> and SBP<sub>brach</sub> were measured simultaneously by a new non-invasive occlusive oscillometric method (Arteriograph, TensioMed Ltd., Hungary) in a healthy population aged 3–18 years with normal BMI (1802 boys, 1572 girls). The diagnosis of JH based on ECG, echocardiography, abdominal sonography, blood-chemical examination, 24-hour-ABPM and arteriography. SBP<sub>ao</sub> and SBP<sub>brach</sub> were measured in JH patients (173 boys, 44 girls) and compared with sex- and age matched control groups. Results were analyzed by Student's t-test.

**Results:** The physiological changes of SBP<sub>ao</sub> and SBP<sub>brach</sub> measured in healthy population are shown in Table 1 and 2. The SBP<sub>brach</sub> increased with age in both genders. The SBP<sub>ao</sub> was remarkably lower in both genders, and the increasing with age was smaller. The SBP<sub>brach</sub> was 147.8 ± 12.1 mmHg in JH boys and 121.7 ± 8.8 mmHg in controls (p < 0.001), while 147.3 ± 15.0 mmHg in JH girls and 117.9 ± 6.9 mmHg in controls (p < 0.001). The SBP<sub>ao</sub> was significantly lower in all groups than SBP<sub>brach</sub> was: 127.4 ± 10.4 mmHg in JH boys, 106.1 ± 7.3 mmHg in control group (p < 0.001), 129.4 ± 14.4 mmHg in JH girls, 104.6 ± 6.4 mmHg in control groups (p < 0.001).

**Conclusions:** First we described the relationship between the SBP<sub>ao</sub> and SBP<sub>brach</sub> measured simultaneously in such a huge healthy population aged 3–18 years. In patients with JH SBP<sub>ao</sub> was considerably lower than SBP<sub>brach</sub> was. On the basis of our findings we may conclude, that the indication of the anti-hypertensive therapy in JH may be reconsidered, and at least the SBP<sub>ao</sub> should be taken into account.

Boys			
Age (ys)	n	SBP <sub>brach</sub> (mmHg) ± 2SD	SBP <sub>ao</sub> (mmHg) ± 2SD
3	44	103.0 ± 5.5	096.8 ± 7.1
4	53	104.1 ± 7.8	095.5 ± 8.3
5	80	106.4 ± 7.2	097.7 ± 7.8
6	120	104.5 ± 6.8	095.3 ± 6.9
7	85	107.8 ± 7.2	098.2 ± 8.1
8	74	107.0 ± 7.1	096.5 ± 7.3
9	92	109.3 ± 6.9	098.8 ± 7.8
10	81	111.2 ± 7.3	099.9 ± 7.5
11	80	111.5 ± 7.5	100.3 ± 7.0
12	101	115.0 ± 7.5	101.9 ± 6.8
13	169	117.2 ± 7.8	102.5 ± 6.7
14	187	119.5 ± 7.4	104.0 ± 6.8
15	171	121.1 ± 8.0	105.3 ± 7.1
16	162	124.5 ± 8.5	107.8 ± 7.2
17	197	126.3 ± 8.6	109.1 ± 7.4
18	106	128.2 ± 8.3	110.1 ± 7.3

Girls			
Age (ys)	n	SBP <sub>brach</sub> (mmHg) ± 2SD	SBP <sub>ao</sub> (mmHg) ± 2SD
3	35	102.5 ± 5.1	097.5 ± 7.8
4	43	103.3 ± 5.2	096.9 ± 6.9
5	44	103.7 ± 6.3	096.0 ± 7.9
6	68	105.0 ± 6.9	096.3 ± 7.3
7	72	106.4 ± 7.3	097.1 ± 7.1
8	39	107.6 ± 7.3	097.6 ± 7.5
9	64	109.6 ± 6.4	098.3 ± 5.8
10	63	111.1 ± 7.3	100.0 ± 6.5
11	49	112.1 ± 6.8	099.9 ± 6.7
12	85	113.5 ± 7.5	101.3 ± 6.4
13	195	115.1 ± 7.7	101.7 ± 6.9
14	189	117.6 ± 8.2	103.7 ± 7.6
15	181	117.2 ± 7.6	104.0 ± 6.9
16	174	117.8 ± 7.4	104.8 ± 6.8
17	175	117.7 ± 8.0	104.2 ± 7.7
18	96	118.8 ± 8.6	105.0 ± 7.5

**PW4-10****International Trial of Promising New Auditory Training Program for Distinguishing Innocent and Pathological Murmurs**

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Recognition of normal and abnormal heart sounds and murmurs is of key importance in detecting heart disease in children and adults but is poorly performed by physicians and students.

Current teaching methods are often archaic, lack clear objectives and not informed by understanding of sound recognition. Repetition is crucial to auditory recognition but absent in many teaching programs. New approaches are needed.

**Methods:** We studied an auditory training program which emphasizes repetition and is validated for patients with hearing loss. The objective

was to distinguish between innocent and pathological murmurs. No medical background is needed, as students learn to discriminate between sounds as phenomena.

2 groups of student volunteers age 20–31 with normal hearing were studied: 20 nonmedical students and 120 senior Australian medical students. After a pre test of 20 random murmur recordings from patients played by computer, both groups performed a one hour auditory training protocol. Murmurs were randomly presented in groups of four with the subject identifying them as normal or abnormal. There is an increase in auditory task difficulty as the subject progresses with the requirement of up to 6 consecutive correct answers before advancing, as with video games. A post test of 20 random recordings occurred immediately after training and again 2 months later. A control group of 42 medical students had no intervention between pretest and 2 month followup.

**Results:** Non medical students improved their mean scores from 72.9 (55–95%) to 90.4% (70–100%) (P .001) while medical students improved from 76.0 (43–100%) to 92% (70–100%) (P <.002) This improvement declined after 2 months to 80.1% (P <.002) for nonmedical students and for medical students, to 82% (50–100%, 95%) which was a non significant increase over the pre test score (p = 0.1). Controls had no change. Students were enthusiastic about the program.

**Conclusions:** This new auditory training program rapidly teaches students to distinguish innocent and pathological murmurs with 90% accuracy. Medical education is not necessary for success with the program which may be very useful for nurses and other health professionals. Reinforcement teaching will likely be important. A trial with Canadian medical students is currently underway.

**PW4-11****Long term outcome of coronary artery lesions after Kawasaki Disease in children**

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The aims of this study were to describe and assess long term outcome of cardiac lesions after Kawasaki Disease.

**Material and methods:** The medical records of 417 patients referred for KD suspicion since 1988 were retrospectively reviewed.

**Results:** 210 patients met criteria for diagnosis of KD, at the age of 2.7 ± 2.5 years (median 2). Time to diagnosis was 7 ± 4.6 days (median 6 days), time to hospitalization 5.7 ± 4.3 days (median 5 days). Time to first echocardiography was 11.4 ± 7.8 days (median 9 days), shorter in more recent period. Median time to intravenous immunoglobulin administration was 8 days (1 to 39). At initial evaluation, 63.8% were free from cardiac lesions, 23.8% (52 cases) had coronary artery lesions (CAL) (aneurisms : 25, dilatation : 27) and 12.4% had «hyperechogen» coronary arteries. Among CAL, 40 were <5 mm in diameter, 9 were 5–8 mm, and 3 were >8 mm (giant aneurisms): one third localized on one coronary vessel, one third on 2 and one third on all 3 coronary arteries. Echographic pericarditis was found in 31 patients, mitral insufficiency in 20 and aortic insufficiency in 2. All patients recovered, except 1 who died from cardiogenic shock due to ruptured chordae. Coronary lesions resolved in 17 of 52 cases (32.6%) and persisted in 35 (67.4%, i.e. 16.7% of all patients): 14 with aneurisms and 19 with dilatations. No patient developed significant long-term coronary artery stenosis. The incidence of aneurisms was lower over the past decade (7.2%). Children with CAL were more likely to have pericardial effusion

(OR 3.00, CI 1.34–6.72) and valvular regurgitation (OR 2.51, CI 1.22–5.16) at diagnosis. However, in case of CAL absence at first echocardiography, these abnormalities were not predictive of CAL at follow-up. Neither valvular regurgitation, nor systolic dysfunction, nor pericardial effusion was associated with persistence of CAL. Male gender, size of CAL, and resistance to immunoglobulin treatment were independent factors predictive of the persistence of CAL.

**Conclusion:** The occurrence of coronary lesions in KD have lessened over time and long-term cardiac outcome is favourable despite persistent coronary lesions. Children with valvular regurgitation or pericardial effusion should have a careful assessment of coronary status at diagnosis.

#### PW4-12

##### **Malignancy after pediatric heart transplantation: a multi-institutional study**

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**Background:** Post-transplant neoplasias are an important cause of morbidity and can potentially be a fatal complication after pediatric heart transplantation (HT). We sought to better determine the frequency, type of malignancy and time of presentation.

**Methods:** We reviewed data on patients follow up at the Spanish Post-Heart-Transplant Tumor Registry from 1984 to 2009. A total of 201 HT was undertaken in patients aged < 18 years old. The data analyzed in the first stage of the study were the incidence of neoplasia, the clinical type and the median follow-up time of diagnosis after HT.

**Results:** Malignancy was diagnosed in 22 patients (10,9%). Post-transplant lymphoproliferative disorders (PTLD) were found in 16 (72,7%) cases. 2 (9%) patients had skin cancer and 3 (13,6%) other tumors. The median follow-up time for the PTLD group was 7,2 years, for the skin cancer group 14,4 years and for the group with other neoplasia was 5 years.

**Conclusions:** About 11% of pediatric HT recipients develop malignancy being PTLD the most frequent (>70%). Further studies should analyze the primary EBV infection, antiviral prophylaxis, the induction treatment and the effects of the new immunosuppressors on the development of PTLD.

#### P-1

##### **Effect of right bundle branch block on T peak to end interval**

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**Introduction:** The interval between the peak and end of the T wave (TpTe) is an index of spatial dispersion in transmural cardiac repolarization (SDR). Children post cardiac surgery with right bundle branch block (RBBB) may have an altered SDR. Patients with right bundle branch block are purported to be at higher risk for arrhythmia. The measurement of TpTe in such children is potentially useful in assessment of arrhythmia risk.

We examined the behaviour of TpTe in the presence of right bundle branch block (RBBB) as a potential marker for arrhythmogenesis.

**Methods:** 45 children with RBBB enrolled. In 42, RBBB occurred post cardiac surgery, 3 se underlying cardiomyopathy. Cohort of 400 normal children served as controls. aAdigital 12 lead electrocardiogram (ECG) was recorded (speed of 50 mm/s) and stored.

Parameters measured TpTe, RR, QT, and JT intervals (leads II and V5). For heart rate correction Bazett formula applied (TpTe/root RR), TpTe B; and Fridericia formula (TpTe/RR<sup>1/3</sup>) TpTe F and calculated the TpTe/QT and TpTe/JT. Results compared to normal data (400 children). Descriptive and analytical statistics, significance level p < 0.05.

**Results:** median age patients 7,6 years + 5,9; controls 5,3 years + 5.1. Mean QRS duration in RBBB group 120 ms (range 88–168 ms). TpTe in leads II and V5 was significantly longer in RBBB patients compared to controls. TpTeB, and TpTeF in leads II and V5 significantly longer than controls. The TpTe/JT (leads II and V5) higher in RBBB patients than controls (this ratio utilised to eliminate potential effects of QRS prolongation on TpTe/QT). All measured values in RBBB patients were significantly different from controls p < 0.05. No patients have thus far presented with life threatening arrhythmias.

**Discussion:** The TpTe reflects global myocardial repolarization and is a surrogate diagnostic parameter. In our cohort of RBBB patients, TpTe and ratio to JT and QT were found to be prolonged when compared to control subjects. No patients to date have developed important ventricular arrhythmia. The role of right ventricular size and haemodynamics on TpTe is under investigation.

#### P-2

##### **Home Monitoring in children and young patients with a congenital heart disease and electronic devices**

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**Aim:** In children and young adults self-perception and self-responsibility is not fully developed. An automated cardiac monitoring system can assist to early diagnose clinical problems or to anticipate device failure in the seriously affected patients with congenital heart diseases (CHD).

**Population:** Out of 150 patients 45 patients received a pacemaker (PM) or defibrillator (ICD) with the Home Monitoring (HM) option (Biotronik, Berlin, Germany). Patients' age at implantation ranged from 5 weeks to 37.6 years [median 12.4 years]. The individual follow up time from the daily monitoring data is 7 days up to 5.3 years (mean 1.7 years).

**Results:** The evaluation-period summarizes up to 80 patient years with successful transmission on 72% of the days, depending on the device type, with 666 clinical relevant event messages received. The messages can be grouped into patient/system related and emergency/follow-up messages. 17% of all messages were categorized as 'emergency' with the need to immediate react to the incoming data. Consequences were system or lead revisions, electro-physiologic studies, reprogramming of parameters, modifications in medication and sports or to further observe. Transmitted intracardiac electrograms (IEGM) reflect



the proper function of the system as well as the actual cardiac electric performance. In 14 patients with an ICD, tachycardia with the need to treat was found in 7 patients. 5 patients had 19 episodes with anti-tachycardic pacing (ATP).

**Conclusion:** The day to day transmission of data routinely and continuously monitored in every PM or ICD markedly improves safety and reliability of electronic device therapy in young patients. High transfer rates increase the probability for early event detection and offer the chance for early intervention. Despite some impact on our clinical workload and legal aspects regarding liability and organization of procedural steps, this system improves therapy in our most critical patients.

### P-3

#### **Long-term Fate of Patients With Congenital Heart Disease Undergoing Cardiac Resynchronization Therapy**

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**Objectives:** CRT is rarely used in pts with congenital heart disease (CHD) and follow-up in available studies is too short. We sought to evaluate long-term impact of CRT in pts with CHD.

**Methods:** 33 consecutive pts with structural CHD (N = 32) or congenital AV block (N = 1) aged median 13 (IQR 8–23) yrs were followed-up after primary (N = 9) CRT-P (N = 29) or CRT-P/D (N = 4) implantation or upgrade from conventional pacing (N = 24) for >6 (median 50, IQR 38–72) mos. Procedures were performed for treatment of dyssynchronous heart failure (HF, N = 29) or to prevent systemic ventricular desynchronization in case of a bradycardic pacemaker indication (N = 4) and associated with additional cardiac surgery in 12 pts. CRT response was defined as improvement of systemic ventricular EF or fractional area of change (FAC) by >10 points and improved or unchanged NYHA class at the end of follow-up.

**Results:** There were 6 adverse outcomes (18.2%) with an actuarial probability of an uneventful therapy continuation at 5 yrs of 77.0%: 1 HF related and 1 sudden cardiac death, 1 heart transplant, 1 pat suffering from neurological damage due to exit block and asystole and 2 therapy terminations because of lead fracture and infection, resp. Among 29 pts treated for dyssynchronous HF (initial median EF or FAC = 23, IQR 16–31%, mean NYHA = 2.3) long-term CRT response was observed in 8/12 pts with systemic LV and 5/17 pts with systemic RV or single ventricle, P = 0.07. Decrease in NYHA class and in the z-score of systemic ventricular end-diastolic dimension was larger in pts with systemic LV (P = 0.038 and 0.006, resp.). 8 surgical CRT system revisions had to be performed in 7 pts (generator replacement for ERI in 6/8) with a reintervention-free survival probability at 5 yrs of 81.6%.

**Conclusions:** Long-term CRT led to sustained improvement of systemic ventricular function in ~2/3 of pts with systemic LV and ~1/3 of pts with systemic RV or single ventricle. It was associated with a significant incidence of heart failure-related adverse events and device complications, resp. Surgical revisions were mainly due to battery depletion. (Supported by the research project of University Hospital Motol MZOFNM2005).

### P-4

#### **18 Years of Paediatric Catheter Ablation in One Country: Long-Term Results**

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**Objectives:** To evaluate long-term results of pediatric catheter RF ablation in one country.

**Methods:** From 1993 to 2010 a total of 695 pediatric ablation procedures were performed in 625 consecutive pts <18 yrs (median age at first ablation 14.8, interquartile range (IQR) 12.4–16.5 yrs) in 3 centers in the Czech Republic to treat 701 different arrhythmogenic substrates (accessory pathway = 427, AVNRT = 203, Mahaim = 19, focal atrial tachycardia = 16, ventricular tachycardia = 16, atrial flutter = 13, incisional atrial tachycardia = 4, twin AV nodes = 3). Structural congenital heart disease was present in 43 pts (7.5%). Indications were patient preference (76.6%), drug refractoriness (14.6%), malignant arrhythmias (6.2%), other (2.1%). Antiarrhythmic drugs were administered in 44.2% of pts. Median follow-up was 13.6 (IQR 4.3–21.1) mos.

**Results:** Acute/long-term success of the primary procedure was 89.1/73.4% for all substrates (AVNRT 98.2/80.3%, accessory pathways 86.9/74.9%, Mahaim 90.0/58.8%, focal atrial tachycardia 61.1/46.2%, ventricular tachycardia 80.0/27.3%, atrial flutter 71.4/58.3%, incisional atrial tachycardia 50.0/25.0%, twin AV nodes 75.0/75.0%). Re-ablation was performed in 74/170 substrates after a primary unsuccessful procedure resulting in long-term cumulative efficacy of 81.1% (533 of 657 attempted substrates). Between 1993–2005 and 2006–2010 median procedure and fluoroscopy time decreased from 154 to 105 and from 24 to 11 min., resp. (P < 0.001 for both). Routine use of non-fluoroscopic navigation (LocaLisa<sup>®</sup>, Medtronic Inc.) in one of the centres from 2010 (36 procedures) carried further decrease in median fluoroscopy time from 14 (period 2006–2010) to 4 min (P < 0.001). Serious complications occurred in 9 pts (1.4%): 3rd degree AV block in 3 (2/203 pts with AVNRT [1,0%], 1 pat with septal pathway), neurological complication in 2, pseudoaneurysm and rupture of femoral artery in 3 and 1 pat, resp.

**Conclusions:** RF catheter ablation was a safe method of arrhythmia treatment in children with long-term efficacy approaching 80%. Patient choice was the most common indication. Procedure and fluoroscopy time decreased with increasing experience and X-ray exposure may further be significantly limited using non-fluoroscopy navigation. (Supported by the research project of Univ. Hosp. Motol No. MZOFNM2005).

### P-5

#### **Permanent Epicardial Pacing in Children: Long-Term Results and Factors Modifying Outcome**

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**Objectives:** We sought to evaluate the results of permanent epicardial pacing in children with respect to risk factors modifying long-term outcome.

**Methods:** All consecutive pts from one country (N = 119, period 1977–2009) undergoing permanent epicardial pacemaker implantation below 18 yrs of age (median 1.8, inter-quartile range (IQR) 0.3–6.4 yrs) were retrospectively studied. A total of

207 pulse generators, 89 atrial and 153 ventricular pacing leads were implanted with a median patient follow up of 6.4 (IQR 2.9–11.1) yrs. Atrioventricular block was the prevailing indication (86.6%). Structural congenital heart disease was present in 76.5%. Pacing system dysfunction was defined by any of the following endpoints: generator and/or lead replacement/revision/abandonment due to exit block, major increase in pacing threshold, fracture or insulation break, patient outgrowth, infection and premature (<3 yrs) battery depletion.

**Results:** Probability of absence of pacing system dysfunction was 79.0/52.1% at 5/10 yrs after implantation. Probability of continued epicardial pacing was 92.8/76.1/58.2% at 5/10/15 yrs and increased in recent implantation era (2000–2009, HR 4.17, CI 1.15–16.67,  $P = 0.030$ ). The use of steroid-eluting leads decreased the risk of exit block (HR 0.25, CI 0.11–0.55,  $P < 0.001$ ) with an actuarial probability of exit block absence at 5 yrs of 95.3% as compared to 76.2% in non-steroid leads ( $P < 0.001$ ). The use of bipolar Medtronic 4968 leads significantly reduced the risk for surgical reintervention because of fracture, insulation break or outgrowth as compared to the unipolar 4965 lead design (96.6% vs 84.2% at 5 years; HR 0.19, CI 0.07–0.46,  $P < 0.001$  for both). The use of the AutoCapture™ feature (HR 14.29, CI 2.94–50.00,  $P < 0.001$ ) and steroid-eluting leads (HR 3.70, CI 1.22–11.11,  $P = 0.020$ ) significantly increased battery longevity. No patient has died because of pacing system failure/infection.

**Conclusions:** The probability of continued epicardial pacing in children was as high as ~ 75/60% at 10/15 yrs after implantation and allowed to defer transvenous pacing to a significantly higher age. The use of bipolar steroid-eluting leads and of the AutoCapture™ feature significantly increased pacing system longevity and decreased the need for surgical re-interventions.

#### P-6

##### **Assessment of electrical and mechanical heart function in children after atrioventricular node slow pathway ablation**

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**Introduction:** Alterations of heart rate (HR), the presence of persistent, inappropriate sinus tachycardia particularly after atrioventricular (AV) node slow pathway ablation have been reported in a variable percentage of patients. It has been suggested that disturbances of autonomic tone may be a contributing factor.

**Purpose:** To evaluate the changes of the conductive system of the heart, autonomic dysfunction, echocardiographic parameters in children late after atrioventricular node slow pathway ablation.

**Methods:** 22 children, who underwent radiofrequency ablation of slow pathway in mean 3,24 years ago, were enrolled into the study. 24-hour Holter recording, electrophysiological transesophageal examination, 2D echocardiography was performed.

**Results:** Sinus cycle length shortened from  $736.6 \pm 134.0$  ms in preablation state in comparison with  $644.4 \pm 179.5$  ms at the late follow-up,  $p < 0.05$ . Mean HR increased from  $72.0 \pm 9.23$  bpm to  $78.1 \pm 5.3$  bpm,  $p = 0.002$ ; maximal HR increased from  $132.5 \pm 16.6$  bpm to  $143.9 \pm 13.0$  bpm,  $p = 0.008$ . The analysis of heart rhythm variability revealed reduction of pNN50 (the percentage of the successive normal sinus RR intervals  $> 50$  ms (%)), from  $26.2 \pm 8.2\%$  to  $20.6 \pm 6.8\%$ ,  $p = 0.01$ ; rMSSD (root mean square of the successive normal sinus RR interval difference (ms)), from  $51.2 \pm 8.2$  ms to  $42.6 \pm 12.6$  ms,  $p < 0.03$ ; and HFC (high frequency component) from  $1014.6 \text{ ms}^2$  to  $706.2 \text{ ms}^2$ ,  $p = 0.007$ , in children, who underwent radiofrequency ablation of slow pathway. There were no significant differences in echocardiographic left and right ventricular parameters except left and right atrial

volumes and their indices that were significantly higher at the late follow-up: left atrial volume increased from  $28.0 \pm 8.7$  ml to  $37.3 \pm 13.9$  ml,  $p = 0.024$ ; left atrial volume index increased from  $15.3 \pm 5.3 \text{ ml/m}^2$  to  $25.1 \pm 8.4 \text{ ml/m}^2$ ,  $p = 0.002$ ; right atrial volume increased from  $16.2 \pm 4.7$  ml to  $22.6 \pm 7.4$  ml,  $p = 0.047$ ; right atrial volume index increased from  $16.3 \pm 5.6 \text{ ml/m}^2$  to  $20.8 \pm 9.5 \text{ ml/m}^2$ ,  $p = 0.05$ .

**Conclusions:** The changes in heart rate, heart rhythm variability, increased atrial volumes and volume indices were revealed in children who underwent radiofrequency ablation of slow pathway.

#### P-7

##### **Selective-site pacing in children using the SelectSecure System: effect on left ventricular function**

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**Introduction:** Few data are available about selective-site pacing in children. A small lead potentially ideal for transvenous pacing in young patients is now available. In order to put in evidence a possible deterioration of left ventricular function, we prospectively evaluated the effect of right ventricular mid-septum (RVMS) pacing on Left Ventricular Ejection Fraction (LVEF) of children undergoing selective-site pacing by SelectSecure Lead System (SSLS).

**Patients and methods:** From June 2006 to January 2011, 50 leads (25 atrial, 25 ventricular) were implanted in 25 patients (10 females) with complete atrio-ventricular block. Mean age at implantation was 9 years (range 3–17), mean weight 31 Kg (range 13–54). All patients received a dual chambers DDD pacemaker. LVEF was evaluated by echocardiography using the Simpson's method at 1, 3, and every 6 months after pacemaker implantation.

**Results:** Median length of follow-up was 24 months (range 6–55). LVEF at first implantation and at last follow-up was  $57.32 \pm 14.7$  and  $61.36 \pm 9.9\%$  ( $p = 0.03$ ), respectively. Deterioration of LV function never occurred. In two patients with heart failure clinical condition and LVEF rapidly improved; in one diuretics and ACE inhibitors were discontinued and he was removed from the heart transplant list; in the second one normalization of cardiac size was observed. Mean QRS duration during spontaneous rhythm and RVMS pacing was  $96.8 \pm 21.5$  ms and  $100.48 \pm 18.1$  ms respectively ( $p = 0.26$ ).

**Conclusion:** RVMS pacing might restore a physiologic electrical activation and synchronization. SSLS is a promising system for permanent intracardiac pacing in children. In our population LV function significantly improved, and particularly so in patients with severe ventricular dysfunction. Although our preliminary results are promising, controlled studies are mandatory to confirm the positive effect of this technique on LV function.

#### P-8

##### **A frequent anomalous morphological pattern of the coronary sinus in corrected transposition of the great arteries: Implications for cardiac resynchronization therapy**

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**Introduction:** Patients with corrected transposition of the great arteries (CTGA) usually present with complete atrioventricular (AV) block and cardiac failure in the late evolution, occasionally requiring resynchronization therapy. We sought to evaluate the

anatomy of the coronary sinus in hearts with CTGA in order to provide morphological guidance to the electrophysiologist to pace the left-sided systemic right ventricle.

**Methods:** A total of 16 hearts from our anatomical collection were analyzed: 8 controls from patients who died of other causes and 8 with CTGA. Mean ages were respectively 61.1 and 15.8 years. The coronary sinus (CS), the oblique vein of the left atrium and the great cardiac vein were dissected and the course of the CS in the left AV groove inspected. The linear distance between the ostium of the CS and the point of drainage of the left atrial oblique vein was measured and normalized by the largest internal diameter of the right AV valve (morphologically tricuspid in control hearts and mitral in corrected transposition).

**Results:** Five of the 8 hearts (62.5%) with CTGA showed the coronary sinus deviated from its normal course in the AV groove and ascending obliquely on the surface of the posterior wall of the left atrium to meet the oblique vein. The maximal deviation distance coincided in all hearts with the point where the left oblique vein (or the persistent left superior vena cava in one heart) reached the coronary sinus. This deviation distance, normalized by the largest diameter of the right AV valve ranged from 0.5 to 0.7. The linear measurement of the ascending portion of the CS in CTGA hearts correlated positively with the largest diameter of the morphologically tricuspid valve ( $R^2 = 0.88$ ,  $p = 0.02$ ). Compared to the controls, in the hearts with CTGA the left oblique vein reached the CS significantly closer to its ostium in the right atrium ( $p < 0.001$ ).

**Conclusion:** The distorted course of the CS and morphology of the left atrial oblique vein in CTGA may impose limitations during CS cannulation and should be taken into account when electrophysiological procedures are considered in this group of patients.

#### P-9

##### **Electrocardiographic screening of one-month-old infants for long QT syndrome in Japan**

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**Introduction:** Electrocardiographic and molecular studies have clarified an association between sudden infant death syndrome and long QT syndrome (LQTS). However, only one prospective study had been conducted in Italy. Our preliminary data showed that the QT interval was longest when infants were 6–11 weeks of age. In Japan, medical examinations for infants at 1 month of age are mandatory. In 2010, we began a prospective study, obtaining electrocardiograms (ECGs) from more than 5,000 subjects at the time of their 1-month examination.

**Methods:** Resting ECGs were recorded in infants from eight areas in Japan. The QT intervals of three consecutive beats were measured. A formula to minimize the effect of heart rate for

infants was used:  $QTc = QT/RR^{0.43}$ . A provisional criterion of  $QTc \geq 0.44$  s was used. To assess the validity of the criterion, infants with a  $QTc \geq 0.43$  s were followed in each area; infants with  $QTc \geq 0.42$  s were followed in the Kagoshima area. ( $QTc$  values by Bazett formula were expressed as seconds<sup>0.5</sup> for reference.) Genetic testing was performed on clinically diagnosed infants.

**Results:** Up to December 1, 2010, subjects were 2,019 infants (age  $31 \pm 3$  days). Mean  $QTc$  was  $385 \pm 18$  ms ( $412 \pm 19$  ms<sup>0.5</sup>). Among them, 20 (1.0%) had a  $QTc$  of  $\geq 0.42$  s. In the Kagoshima area, 16 of 1,230 (1.3%) had the same values and all 16 infants had a  $QTc$  of  $< 0.42$  s a few weeks later. Six (0.3%) had a  $QTc$  of  $\geq 0.43$  s; all were followed. Of the six, one (431 ms or 458 ms<sup>0.5</sup> at one month) was started on treatment at 51 days (500 ms or 535 ms<sup>0.5</sup> on the same day). Only one infant met the provisional criterion (463 ms or 492 ms<sup>0.5</sup> at one month) and treatment was started at 60 days (496 ms or 524 ms<sup>0.5</sup>). Propranolol and mexiletine were administered. Genetic testing revealed a *KCNH2* mutation (3065 T deletion) in the second case. The other is under investigation.

**Conclusion:** The incidence of LQTS infants who are clinically diagnosed may be close to 1:1,000. ECG screening for LQTS in infants may be effective; cost-effectiveness of the screening should be further investigated.

#### P-10

##### **Isosorbide Dinitrate Is a Safe Provocative Agent In Head up Tilt Test in Children and Adolescents**

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**Introduction:** Neurally mediated syncope (NMS) in the children and adolescents is the most likely cause of syncope. Head up tilt test (HUT) is used in diagnosing children and adolescents with NMS. Conventional HUT test protocol is time-consuming and the sensitivity of this method is relatively low. Pharmacologic provocative agents such as nitroglycerin and isosorbide dinitrate (ISDN) are commonly used to increase diagnostic yield of HUT test. It has been reported that sublingual ISDN-HUT test is suitable for routine clinical practice in children and adolescents with unexpected syncope. But in that protocol, the total tilt duration is too long for children to tolerate. This study evaluated the advantage and safety of a short ISDN-HUT test protocol for diagnosing NMS in children and adolescents.

**Material and Methods:** We studied 172 patients (84 males and 88 females) referred with unexplained syncope. In case group 43 patients had mean ( $\pm$ SD) age of  $11.8 \pm 3.7$  years and in control group 129 patients had mean ( $\pm$ SD) age of  $11.7 \pm 3.7$  years. The patients in case group were tilted to an angle of 65 degrees for 20 minutes (unmedicated phase). If test became negative the patient would receive 0.1 mg/kg ISDN sublingually in supine position. After 5 minutes, the table was tilted to an angle of 65 degrees for a maximum of 20 minutes or until the test became positive (medicated phase). The patients in control group were tilted to an angle of 65 degrees for 40 minutes (conventional HUT test protocol).

**Results:** Overall 36 patients (83.5%) in case group 17 patients (39.5%) in unmedicated phase and 19 patients (44%) in medicated phase and 80 patients (62%) in control group had a positive response ( $P = 0.009$ ). All patients in the case group tolerated ISDN very well.

**Discussion:** Our finding showed that short ISDN-HUT test protocol for diagnosing NMS is safe and has a higher rate of positive response than conventional HUT test protocol.



**P-11****Fifteen year experience with accessory pathway ablation in children and young adults with congenital heart disease by a single pediatric electrophysiology team**

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**Introduction:** Catheter ablation (CA) of accessory pathways (APs) has evolved from a procedure usually performed after failure of medical therapy to the main therapeutic approach in children and congenital heart disease (CHD) patients. The purpose of this study was to report our experience accumulated over the past 15 years with CA of APs in children and CHD patients.

**Methods:** Review of a database of consecutive CA procedures performed by a single pediatric electrophysiology team.

**Results:** Two-hundred and twenty nine patients aged 0.28–35.85 years ( $10.97 \pm 4.05$ ), underwent CA of AP; 31 pts had CHD (13.5%), the most common being Ebstein's anomaly (12 pts). Five pts had complex CHD. Forty-two pts (18.3%) had asymptomatic preexcitation, 10 (4.4%) had permanent junctional reciprocating tachycardia (PJRT), 3 (1.3%) had Mahaim fibers and the remaining (75.9%) had orthodromic reciprocating tachycardia. Acute success was achieved in 96.5% and recurrence was observed in 13.1% of patients (7 left, 16 septal and 8 right-sided), including 3 of 12 with Ebstein's anomaly. After repeat ablation procedures (1–3) in 28 pts, only 4 pts remained with a functioning AP (final success rate: 98.2%). Fluoroscopy time decreased from a mean of  $31.30 \pm 27.98$  min to a mean of  $8.54 \pm 9.45$  min after the introduction of a system of non-fluoroscopic navigation (NavXTM, St Jude Medical). Complications occurred in 12 pts (5.2%), but only 3 were considered significant, including 1 patient each with aortic insufficiency, mitral insufficiency, and AV block lasting for 5 days, and none requiring intervention.

**Conclusions:** Catheter ablation has evolved to an extremely efficacious and safe therapeutic approach for the treatment of arrhythmias related to accessory pathways in pediatric and CHD pts. Despite the difficulty imposed by certain types of CHD, equal long-term success can be achieved in these patients as well as in those with structurally normal hearts.

**P-12****Sotalol in Foetal Atrial Tachyarrhythmias.****A Retrospective Analysis**

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**Objective:** Investigating the efficacy and side-effects of Sotalol as a first-line treatment in foetuses with atrial tachyarrhythmias.

**Methods:** From 2004 to 2010 thirty foetuses with atrial tachycardia – AV-node re-entry tachycardia (SVT) and atrial flutter (AF) – were retrospectively evaluated. The heart rate, presence of hydrops, antiarrhythmic therapy, success rate and time to conversion to sinus rhythm (SR), relapse, side effects and maternal QTc interval and postnatal outcome were analysed.

**Results:** In 20 of the 30 foetuses SVT was present (200–300 bpm). In 10 foetuses AF was present with atrial rates of 300 to 470 bpm and a mean ventricular rate of 205 bpm.

Nonhydropic foetuses ( $n = 22$ ) received Sotalol as initial therapy. Of the eight hydropic foetuses, six received Sotalol and two Flecainide as initial treatment. In three cases Digoxin and in two Flecainide was added.

28 of the 30 foetuses converted antenatally to SR with a mean duration to conversion of seven days. Relapse occurred in three, but was solved. In two nonhydropic foetuses with AF, treated with Sotalol and Digoxin, only rate control was achieved. There were no deaths.

Minor maternal adverse effects were encountered. Two mothers had a QTc interval of  $> 470$  ms (487 and 497 ms); Sotalol was continued.

After birth, in all foetuses but two SR was confirmed. Antiarrhythmic therapy was discontinued. Two had heart rate controlled AF. However, during the newborn period, rhythm disturbances were seen in 10 of the 30 infants.

In one hydropic foetus, presenting at 36 + 5 weeks with SVT (280 bpm) and born with SR at 37 + 1 weeks, an approximately seven weeks old cerebral infarction was seen on MRI. All others showed normal neurological development.

**Conclusion:** Sotalol and the combination of sotalol with flecainide were very successful in foetal atrial tachyarrhythmias. In the SVT-group conversion rates reached 100%. The conversion rate in the AF-group was 80%, with rate control in the remaining 20%. Sotalol should be considered as a drug of first choice in the treatment of foetal SVT and AF. Flecainide should be considered as a second-line drug in cases of therapy resistant atrial tachyarrhythmias.

**P-13****Outcomes of Radiofrequency Ablation and Cryoablation in Children and Adolescents**

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**Introduction:** Radiofrequency ablation (RF) and cryoablation are now accepted as routine treatments in older children and adolescents. However, data from the current era is limited particularly in relation to cryoablation in children. We aimed to review the safety and efficacy of our approach. We primarily use RF and reserve cryoablation for locations where there is an increased risk of heart block.

**Methods:** Data from 126 consecutive cases undertaken at a regional children's hospital between January 2006 and January 2010 was studied. All the procedures were carried out under general anaesthesia.

**Results:** The patients' ages ranged from 5 to 18 years (mean 13.8 years, SD  $\pm 2.4$ ). Their weights ranged from 16 to 94 kg (mean 53.8 kg, SD  $\pm 14.5$ ), 61 (54%) were male. The follow up period was between 6 to 54 months (median 26). Indication included focal atrial tachycardia in 3, Right ventricular outflow tract ventricular tachycardia in 1, Fascicular ventricular tachycardia in 2, Atrioventricular nodal re-entrant tachycardia in 48, Wolff Parkinson White syndrome in 63. RF was used in 117 patients, 2 of these patients had congenital heart disease and electro-anatomic navigation was used in both. On table success was seen in 95% (111/117). Recurrence was seen in 8% (9/111). Repeat procedures were performed in 7 patients. At a minimum 6 month follow up and including repeat procedures overall success with RF was 93% (108/117). Cryoablation was used in 9 patients, all of whom had Anteroseptal accessory pathways.

On table success was seen in 66% (6/9). Following repeat procedures success was seen in all expect 1 patient. In this patient there was temporary heart block secondary to cryocatheter pressure. Overall success for cryoablation was therefore 77% (8/9) at minimum six month follow up. Screening time per case was 5–48 minutes (mean 14.2, SD  $\pm$ 9.4), there was no significant difference between RF and cryoablation. There was no permanent heart block and morbidity was limited to a skin burn in 1 patient.

**Conclusion:** Catheter ablation in children and adolescents appears to be safe. An approach incorporating cryoablation further improves safety particularly in ablation proximal to native conduction tissue.

#### P-14

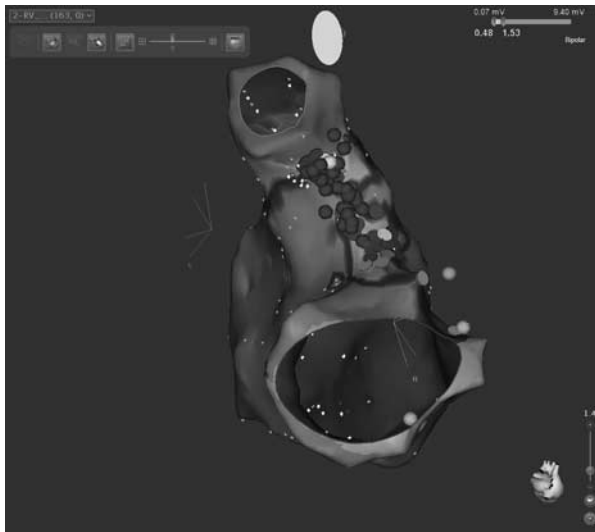
##### **Identification of electrophysiological substrate in patients with repaired tetralogy of Fallot.-**

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**Introduction:** Patients with repaired tetralogy of Fallot (TOF) represent a new category of patients referred to electrophysiology laboratory for ventricular arrhythmia (VA) mapping and ablation. Different anatomical regions have been identified as potentially responsible for reentry: ventricular septal defect (VSD) patch, surgical incisions, right ventricular outflow tract (RVOT) patch. We aimed to investigate electrophysiological substrate responsible for potential VA in patients with repaired TOF.

**Methods:** All patients with repaired TOF referred to the CHU de Bordeaux for VA evaluation from January 2008 to april 2010 underwent right ventricular (RV) 3D. Sinus activation and voltage mapping was then performed before VA induction  $\pm$  ablation.



**Results:** 7 patients (4 male,  $42.5 \pm 12$  years old) underwent RV mapping during VA evaluation. Surgical repair of TOF had been realized  $36 \pm 11$  years before the procedure. All patients displayed a right bundle branch block on 12 lead electrocardiogram.

Sinus rhythm RV activation begins in all patients in the septum and then activates the RV centrifugally with a zone of slow conduction with a double potential ( $100 \pm 30$  ms) going from the tricuspid annulus (TA) to the RVOT. Voltage maps (figure) show systematically the same pattern of a zone of low voltage ( $<1.5$  mV) due to the VSD repair close to the RVOT scar area. This area fits with the slow conduction area. In the 2 patients

with sustained ventricular tachycardia (VT), critical isthmus was located in this area.

**Conclusions:** Specific activation and voltage pattern was found in these Fallot patients. In the 2 patients with sustained VT, the critical isthmus was found between VSD repair patch and RVOT scar.

#### P-15

##### **Heart rate variability in young patients with hypertrophic cardiomyopathy is a useful indicator of risk for complex ventricular arrhythmias**

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**Background:** Heart rate variability (HRV) is an accepted tool for studying imbalance in the autonomic system non-invasively. Alterations in HRV indicating increased activity of sympathetic nervous system has been recognized as a risk factor for cardiovascular mortality, and been suggested to predispose to ventricular arrhythmias in adult patients with hypertrophic cardiomyopathy (HCM). We have used this methodology to study autonomic nervous system activity in young symptom-free patients with familial HCM.

**Methods:** HRV was studied by a 5 min resting ECG-recording analysed by commercial software (CardioPerfect). Initial analysis of a control group of 51 subjects with echocardiographically normal hearts in an age-range between 1–29 years of age indicated a positive correlation between age and the low/high-frequency (LF/HF) component ratio ( $p = 0.0007$ , correlation coefficient 0.46). Between the ages of 10–21 there was also a significant gender difference in the LF/HF component ratio, males 0.99 [95% CI 0.31–1.67] as compared to 0.57 [0.43–0.75] in females. We identified 23 young patients with asymptomatic HCM, who had had HRV analysis performed before any medical therapy had started (mean age 11.3 years, range 1.9–22), and compared those with age- and gender-matched controls.

**Results:** The groups were well age-matched (mean age controls = 11.2; Mann-Witney  $p = 0.83$ ). HCM-patients had a significantly higher LF/HF ratio,  $1.25 \pm 0.96$  (mean  $\pm$  SD) as compared to  $0.53 \pm 0.24$  in controls ( $p = 0.001$ ). The alteration was caused both by a significantly higher normalized LF ( $49.3 \pm 16.2$  versus  $33.2 \pm 10.1$ ;  $p = 0.001$ ) and a significantly lower normalized HF ( $50.7 \pm 16.2$  versus  $66.8 \pm 10.1$ ;  $p = 0.001$ ) in the HCM-patients. All HCM-patients had 24h ECG-monitoring performed. 2 patients had identified ventricular tachycardia on 24h ECG, both had LH/HF ratio  $>2.0$ . Among the patients without identified complex ventricular arrhythmia only 2/21 had an LH/FH ratio  $>2.0$  ( $p = 0.02$  Fischer's exact test). In addition, the two that had a high LF/HF ratio without ventricular arrhythmias both had first-degree relatives with identified ventricular tachycardia or resuscitated cardiac arrest.

**Conclusions:** Even asymptomatic patients with HCM diagnosed in childhood and adolescence have alterations of autonomic nervous balance suggesting an increased activity of the cardiac sympathetic system. An LF/HF ratio  $>2.0$  is a marker of increased risk of sustained ventricular arrhythmia in this age-group.

#### P-16

##### **Cardiac resynchronization therapy in children – experience of the Hungarian Paediatric Heart Centre**

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**Introduction:** Cardiac resynchronization therapy (CRT) is an accepted non-pharmacological treatment of heart failure, and an

option for bridge-to-transplantation in adults and children. Though, CRT is used in different diseases; mode of implantation and outcomes can vary, respectively.

**Method:** Clinical conditions, mode of implantation and complications were recorded in patients treated with CRT between 18.10.2004–01.01.2011 in our Institute. Impact of CRT on long-term clinical response was evaluated.

**Results:** Left ventricular CRT was performed in 12 and right ventricular CRT in 1 patient with congenitally corrected transposition of the great arteries, respectively. Mean age at CRT: 3.75 years. Male-to-female ratio: 10:3. Mean follow-up period: 3.6 years.

**Clinical conditions:** Congenital atrioventricular block (congAVB) with ventricular dysfunction developed during pacemaker (PM) therapy (6); congAVB with ventricular dysfunction present at time of PM-implantation and progressive heart failure (2); postoperative AVB (popAVB) with ventricular dysfunction developed during PM therapy (2); popAVB with ventricular dysfunction present at time of PM-implantation and progressive heart failure (1); hereditary DCM (1); myocarditis (1).

**Mode of implantation:** Epicardial electrodes (9), endocardial electrodes (1), dual-chamber endocardial system up-dated with epicardial left ventricular electrode (2), right ventricular epicardial, right atrial and coronary sinus endocardial electrodes (1).

**Function:** CRT-pacemaker (12), CRT-defibrillation (1).

**Complications:** electrode dislocation (1), decubitus above generator (1), phrenic muscle stimulator (2), postoperative low cardiac output syndrome (2).

**Impact on clinical course:** Cong/popAVB + good systolic function at PM-implantation: improvement (7); CRT discontinued due to phrenic muscle stimulator (1). CongAVB + ventricular dysfunction at PM-implantation: improvement, but heart transplantation (htx) after 2.6 yrs (1); exitus (prae-transplant era) (1). PopAVB + ventricular dysfunction at PM-implantation: no improvement (LVAD 3 months postCRT) + htx (1). DCM: exitus (prae-transplant era) (1). Myocarditis: worsening (1).

**Conclusions:**

1. Ventricular dysfunction related to congAVB after PM therapy is the most common clinical condition, where CRT was performed in paediatric patients.
2. CRT improved ventricular dysfunction developed during PM therapy regardless of the etiology of AVB.
3. CRT was ineffective as a rescue therapy for DCM in the prae-transplant era and in myocarditis, but had role in bridge-to-transplantation in DCM patient with congAVB.
4. In children mostly epicardial leads are used for CRT, and the complication rate is not negligible.

#### P-17

##### **One cryoablation is often enough to treat children with septal arrhythmia substrates in the right atrium**

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**Introduction:** Cryoablation is the preferred method of ablation for children with arrhythmia substrates near the normal conduction system. It provides increased safety compared to radiofrequency energy due to the characteristics of cryoenergy tissue effects, ability to cryomap and firm catheter attachment during cryoenergy application. Freeze-thaw cycles or a line of adjacent lesions have been recommended to decrease recurrence risk, which has been suggested to be higher than with radiofrequency

ablation. However, at least in the smallest children, extra lesions might cause additional unnecessary risks.

**Methods:** Children treated with only 1 or 2 cryoapplications were collected out of 62 consecutive children treated with cryoenergy during Dec 2005 and June 2010. These 25 patients were analyzed for arrhythmia substrates, ablation procedures, and recurrence rate. The median age was 13.5 yrs (range 7.3–18.3, 75% and 25% interquartile 12.5–15.7), weight 52 kg (27–80, iq 42–66) and height 166 cm (131–187, iq 150–175). The tachycardia mechanism was AV nodal re-entrant tachycardia (AVNRT) in 19 patients (76%), accessory pathway in 5 (20%) and atrial ectopic tachycardia originating near coronary sinus ostium in 1 (4%). The accessory pathway location was right anteroseptal in 1, midseptal in 2 and posteroseptal in 2. The cryocatheters used were 7 F Freezor Xtra 3 with a 6 mm tip in 16 pts and Xtra 1 with a smaller curve in 9 pts. The median fluoroscopy time using NaVX navigation system was 5.5 min (1–26, iq 3–9), radiation dose 80  $\mu$ Gym<sup>2</sup> (24–570, iq 35–138) and procedure time 2 hrs (0.8–3.5, iq 1.5–2.5).

**Results:** 18 pts (29% of all pts) were successfully treated with one cryoapplication of 4 minutes or less, with only one recurrence. Additional 7 (11%) patients were cured with 2 lesions. The one recurrence occurred in a posteroseptal accessory pathway and was successfully treated in another procedure. The median follow-up time of the 25 patients was 24 months (5–59, iq 14–38).

**Conclusions:** Up to 40% of arrhythmia substrates in right atrium can be successfully treated with only one or two cryolesions in pediatric patients in straight-forward cases. The recurrence rate was 4% during a median follow-up of 2 years.

#### P-18

##### **Late recovery of surgical atrio-ventricular block is not exceptional**

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**Introduction:** Surgical atrioventricular (AV) block may complicate cardiac congenital surgical procedures. It is generally considered permanent when AV-block persists beyond the 14th postoperative day. We studied the incidence of spontaneous late recovery of AV conduction after surgical AV-block.

**Methods:** We retrospectively reviewed our Pediatric Cardiology database for all cardiac surgical procedures between January 1993 and November 2010 in subjects <18 years old. All patients with 2nd or 3rd degree AV-block persisting beyond the 14th postoperative day were included. Late recovery was defined as recovery of AV conduction to normal or to first degree AV-block >14 days postoperatively.

**Results and discussion:** During the study period, 2808 cardiac surgical procedures on cardiopulmonary bypass were performed. Beyond the 14th postoperative day, 2nd degree AV-block was present in 7 (0.25%) patients and 3rd degree AV-block in 68 (2.4%) patients. Late recovery of 2nd degree AV-block occurred in 3/7 patients (42.9%) after 2 months, 3 years and 6.5 years. Late recovery of 3rd degree AV-block occurred in 3/68 (4.4%) patients after 2 months, 3 months and 7 years.

The surgical procedures causing AV-block in the 6 patients with late recovery of AV conduction-block were: AV septal defect correction with mitral valve plasty in 3 (two 2nd and one 3rd degree AV-block), mitral valve plasty in 1 (2nd degree AV-block) and ventricular septal defect closure in 2 (both 3rd degree AV-block) subjects.



None of the patients with recovered AV conduction relapsed into 2nd or 3rd degree AV-block during the study period. Reprogramming of the pacemaker as a back-up device was possible, and prolonged pacemaker battery longevity.

**Conclusion:** Spontaneous late recovery of AV conduction in patients with surgical AV-block is not exceptional and should be searched for during chronic follow-up. Reprogramming the pacemaker to back-up at low rate prolongs pacemaker longevity. It still needs to be determined if pacing system replacement is indicated in case of exit block or battery depletion.

#### P-19

##### **Low incidence of inappropriate shock in children with implantable cardioverter defibrillator. A single-institution experience**

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**Introduction:** Inappropriate shock of implantable cardioverter defibrillator (ICD) is known to be more frequent in pediatrics, secondary to high incidence of lead failure, sinus or atrial tachycardia and over sensing in this population.

**Methods:** We report a single-institution experience of ICD implantation in children. Between January 2003 and December 2010, 24 ICD implantations were performed at mean age of 11 years (range 4.5 to 16), and mean weight of 37 kilograms (from 19 to 60). Indication was secondary prevention for 14 patients (58%) and primary prevention for 10 (42%). Twelve patients had primary electrical diseases, 9 cardiomyopathies, 2 had prior surgical repair of a congenital cardiac defect and one had cardiac tumor with inducible ventricular fibrillation. Implantation was performed transvenously for 10 patients. The 14 others, fewer than 40 kilograms of weight, had epicardial pace-sense leads, with ICD coil in the pleural space and device placed horizontally under the heart. Majority of the ICD receivers (80%) were on high dosage of beta-blockers. All, but one, had a tachycardia detection rate over 200 beats per minute.

**Results:** At midterm follow-up, mean 28 months ( $\pm 26$  months), only two patients received inappropriate shocks (8%), while 9 received appropriate ICD therapy (37%). Three patients had lead failure: a coil migration, an undersensing epicardial lead and an insulation break. Among those failing leads, one was placed transvenously and 2 by nonstandard approach. There was one intraoperative fatality reported on a 5 years old girl with restrictive cardiomyopathy, and another at adult age after heart transplant.

**Conclusion:** Incidence of inappropriate shocks, and lead failure could be effectively decreased by nonstandard placement in children under 40 kilograms, by beta-blockers medication and by personalized programing as reported in this series.

#### P-20

##### **Contact Force Controlled Zero-Fluoroscopy Catheter Ablation of Right-Sided and Left-Atrial Arrhythmias**

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**Introduction:** In children and young adults, radiation exposure should be avoided whenever possible in order to reduce or

eliminate its long-term malignancy risk. However, the avoidance of fluoroscopy during catheter ablation of cardiac arrhythmias guided by a 3D mapping and navigation system is hampered by possible high contact forces that can occur unintended. Here we present a technique for zero-fluoroscopy catheter ablation of various cardiac arrhythmias employing an ablation catheter with integrated force sensor.

**Methods:** After informed written consent, a 7-French irrigated ablation catheter with integrated force sensor for real-time measurement (10 Hz) of tip electrode-tissue contact force (sensitivity <1 g; TactiCath, Endosense, Geneva, Switzerland) was introduced via the right femoral vein. Using an electro-anatomical mapping system (EnSite NavX, St. Jude Medical, St. Paul, MN, USA) with the "belly" patch surface electrode as reference, geometric contours of the external and common iliac vein, the inferior and superior vena cava (IVC, SVC), right atrium (RA) and coronary sinus (CS) were created by sweeping the catheter tip across the respective structures. Applied maximal and mean forces were below 100 and 50g, respectively. Then, using the formerly created contours, a steerable decapolar catheter was advanced via the right femoral vein into the CS and a quadpolar catheter into the right ventricle. Transseptal access was gained under transesophageal guidance.

**Results:** Zero fluoroscopy catheter ablation was initially performed in 9 adult patients (Arrhythmias: 4 AVNRT, 2 typical atrial flutter, 1 RVOT-non-sustained-VT, 2 WPW with left atrial accessory pathways, 1 symptomatic paroxysmal atrial fibrillation) and then in 5 patients aged 10–18 years (Arrhythmias: 4 AVNRT, 1 WPW with left atrial accessory pathway in a 10-year-old boy). In only one adult patient this approach failed due to an accessory pathway within an aneurysm of the coronary sinus which was visualized by coronary sinus angiography. No procedure-related complications occurred.

**Conclusion:** Zero fluoroscopy catheter ablation of right-sided and left atrial cardiac arrhythmias seems in general to be feasible, effective and safe when a 3D navigation system is combined with real-time tissue-tip contact force measurements.

#### P-21

##### **Congenital Junctional Ectopic Tachycardia – difficult clinical problem**

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**Introduction:** Congenital junctional ectopic tachycardia – JET is a rare tachyarrhythmia, may be a reason of post tachycardia cardiomyopathy, is associated with high morbidity and mortality. The purpose of the study was to present our experience with JET in small children.

**Material and methods:** We have 7 children (5 boys) with congenital JET, in 5 of them the tachycardia was diagnosed and treated (digoxin, sotalol or amiodaron) in their fetal life, in one at neonatal period, and in one at age of near 3 year. Among the patients we have two couples of brother and sister (one of them are twins).

**Results:** At the beginning of observation all children presented incessant tachycardia, they have normal heart with good function, on chest X-ray cardiac index was 0.5–0.55, on ECG all had JET with heart rate 155–300 beats/min. In all children we

started aggressive pharmacotherapy with two or three drugs (digoxin, propranolol and propafenon, sotalol or amiodaron). During the follow-up period (ranged from 7 months to 8 years) in all but one we slowed heart rate on medication. In one boy with very fast JET rate emergent radiofrequency (RF) catheter ablation was necessary followed by epicardial pacemaker system implantation at the 7th month of life, he is still on drugs with JET or pacing rhythm. Three children still have JET, with drug control heart rate, one has sinus rhythm and short periods of JET, one brother and sister (twins) converted to sinus rhythm after 5 years of observation.

**Conclusions:** Young children with congenital JET need aggressive pharmacotherapy, ablation procedure may be necessary very early, in some children sinus rhythm may recover.

## P-22

### Safety and efficacy of simplified femoral approach with Gold Tip Electrodes for RFCA on paroxysmal SVTs in children

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**Background:** Radiofrequency catheter ablation (RFCA) in children has been widely used for treatment of supraventricular arrhythmias (SVT). Gold-tip electrodes, with higher conductivity, less risk of thrombus and charring formation have not been widely evaluated in children. A simplified approach with the use of two catheters has not been reported in pediatric population. The aim of the present study was to evaluate a short and long-term safety and efficacy of RFCA for SVT with minimal approach with Gold-tip ablation electrodes.

**Methods:** The studied group consisted of consecutive patients aged <19 years referred for the first RFCA of SVT. A standard 2-catheter approach with femoral vascular approach was used. A standard decapolar catheter introduced in the coronary sinus and mapping/ablation Gold-tip catheter for dynamic mapping and ablation were used.

**Results:** Between January 2007 and May 2010, 104 children were referred for the first RFCA of SVT. There were 52 cases of atrioventricular nodal reentry tachycardia, 54 cases of accessory pathways or accessory pathways dependent tachycardias including 4 atriofascicular pathways and 5 cases with atrial tachycardias. In 7 cases coincidence of 2 arrhythmias was recognized. Standard simplified 2-catheter femoral approach was performed in 93/104 (90%), but in further 10 procedures its modification was used (additional subclavian vein access (n = 1), 3rd catheter (n = 2), intracardiac echocardiography (n = 2) or single catheter approach (n = 7). A mean number of catheters used simultaneously during mapping and ablation were 1.9 +/- 0.3. In 2 procedures a cross over to Gold cooled tip catheter were used. Total procedure, observation and X-ray exposure time of RF application was 65 +/- 29 min, 18 +/- 10 min and 129 min, respectively. Using a 50–60 Watt/50–60°C generator setting a mean 36 +/- 13 maximal Watt and 54 +/- 7°C maximal temperature were achieved. A mean 15 +/- 15 of RF applications were performed with a mean application time of 340 +/- 272 sec. Intraprocedural

efficacy was 97% without significant complications. Steam pops phenomenon, thrombi and charring formations on tips of ablation catheters were not recorded. No late complications were recorded. **Conclusions:** Simplified femoral approach with Gold-tip electrodes for RFCA is a safe and efficient method for treatment of supraventricular arrhythmias and accessory pathways in children.

## P-23

### Catheterablation of accessory pathways in children and adolescents: results in 167 consecutive patients

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**Introduction:** Catheterablation (CA) has become the treatment of choice for accessory pathway (AP) mediated tachycardia. Despite of the high success rate there are pathways which are not amenable for catheter ablation or need more procedures.

**Methods:** A total of 167 consecutive patients younger than 19 years with ventricular preexcitation or AP mediated tachycardia and normal heart intended to treat with CA in our institute were included. Study period 2004–2010. Ablation energy (radiofrequency: RF, cryo: C) and pathway localisation (right:R, left: L, septal: S) related to success and recurrence were evaluated.

**Results:** A total of 176 APs were diagnosed in 167 pts (R:25, S:81, L:70). CA was abandoned in 15 pts because of high cost benefit ratio (S:15, Cryomapping:11). Acute success was achieved in 142/152 pts, with 151 Aps (R:17, S:62, L:72, RF:133, C:18). 7/10 pts had repeat procedure and in 4/7 success were achieved in the later procedure (R: 2, S:2, RF: 4). 18 pts were lost for follow up. 103 pts (R:10, S:43, L:49, RF:91, C:11) had no recurrence during the follow up (median:1 yr). 17 pts had recurrence of preexcitation (R:3, S: 9, L:5, RF:13, C: 4). 10/17 pts had repeat procedure in all with acute success but in 4 pts with further recurrence (R:1, S:2, L:1). 4 pts ablated with concealed AP and recurrent palpitation had a redo procedure in 3/4 with success (R:2, L:1). Recurrence after RF: R:4/11, S: 6/62, L: 5/63 and after C: R:1/2, S:4/16, respectively.

**Conclusion:** Despite of the safety of cryoenergy significant number of septal APs are not amenable for CA in children. Recurrence rate of septal APs after cryoablation is more than twice as after RF ablation. CA of left sided APs have the highest success rate and lowest recurrence rate. Right sided APs have the highest recurrence rate after CA.

## P-24

### Population-based long term outcome of pediatric catheter ablation of common supraventricular tachycardias in patients without structural heart disease

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**Introduction:** Catheter ablation is the preferred treatment method of common SVT not only in adults, but also in children and adolescents. Long term follow-up data on pediatric patients is still incomplete.

**Methods:** A catheter ablation registry was created in November 1997 when pediatric ablation procedures were started in our institution. A single pediatric centre covers the whole country with a population of 5.4 million people. Based on the registry, long term follow-up data were collected with a questionnaire and from charts with 98% coverage. Patients with congenital heart malformations were excluded from this analysis except those with trivial defects.

**Results:** 296 patients underwent catheter ablation, and 269 (91%) were successfully ablated in 1–3 procedures. 66% had an accessory pathway (AP), 31% atrioventricular nodal reentrant tachycardia (AVNRT) and 3% atrial tachycardia. The success rates were 89%, 93% and 71%, respectively. Serious complications were rare: one patient had complete AV block needing pacemaker and one patient had a haemopericardium needing drainage. Before introducing cryoablation in 2005, five patients had temporary second degree AV conduction block, all of which resolved by time. The median follow-up time was 4.8 years (1.0–12.6 yrs).

The recurrence rate after successful ablation was 7.8% (21 patients). Ten of the recurrences occurred several years after the procedure. In addition, two patients have had atrial fibrillation and one a focal atrial tachycardia as young adults after an ablation procedure as a child; two procedures were performed for AVNRT and one for AP. After ablation procedure, eight patients (3%) complained of higher exercise-induced heart rate than before the procedure.

**Conclusions:** Long term outcome of pediatric catheter ablation of supraventricular tachycardia is excellent with a low complication rate. Recurrence rate was 7.8% in our patients with a median follow up of 4.8 years. Half of the recurrences occurred late, several years after the initial procedure.

#### P-25

##### **The funny current channel HCN<sub>4</sub> delineates the developing cardiac conduction system in the chicken heart**

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**Introduction:** Hyperpolarization-activated cyclic nucleotide-gated channel 4(HCN<sub>4</sub>) in the mouse is expressed in the developing cardiac conduction system (CCS). In the sinoatrial node (SAN), HCN<sub>4</sub> is responsible for the funny current. To date no data is available on HCN<sub>4</sub> expression during chicken CCS development. **Objective:** To determine the full-length sequence of Hcn<sub>4</sub> and describe its expression pattern during development in relation to the CCS in the chicken embryo.

**Methods:** Hcn<sub>4</sub> RNA expression was studied by in situ hybridization in sequential chick developmental stages (HH11–HH35) and immunohistochemical stainings were conducted for the myocardial protein cTnI and the cardiac transcription factor Nkx2.5.

**Results:** We obtained the full-length sequence of Hcn<sub>4</sub> in chick. Hcn<sub>4</sub> expression was observed early in development in the primary heart tube. At later stages, expression became restricted to transitional zones, comprising the sinus venosus myocardium where the SAN develops, the atrioventricular canal myocardium, the primary fold located between the developing ventricles to form the interventricular septum, and the developing outflow tract. These zones are flanked by the atrial and ventricular working myocardium. Further in development, Hcn<sub>4</sub> expression was restricted to the SAN, the atrioventricular node, the common bundle, the bundle branches and the internodal and atrioventricular ring myocardium.

**Conclusion:** We have identified Hcn<sub>4</sub> as a functional marker of the developing CCS in the chick. The primary heart tube expresses Hcn<sub>4</sub>, which is later restricted to the transitional zones and eventually the elements of the mature CCS. Furthermore, we propose that expression patterns during development might

explain the occurrence of arrhythmogenic anatomical predilection sites in adults.

#### P-26

##### **Progression to first degree heart block in preschool children fetally exposed to Ro52 autoantibodies**

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**Introduction:** Isolated congenital complete atrioventricular block (AVB) is associated with transfer of maternal anti-Ro/SSA autoantibodies to the fetus and has a high mortality and morbidity. The importance of first- and second degree AVB developing in the fetus is less well studied, including its long term prognosis. This study evaluated children fetally exposed to maternal anti-SSA/Ro antibodies in terms of signs of impaired atrioventricular (AV) conduction or myocardial disease and correlated prenatal Doppler findings to outcome.

**Methods:** A cohort of 57 children fetally exposed to anti-SSA/Ro was identified and grouped in accordance to (A) prenatal Doppler signs of first-degree AV block (1°AVB) or (B) normal findings, and examined by ECG, 24-h Holter and Echocardiography. Outcome was differences for variables obtained by ECG, Holter and Echocardiography.

**Results:** The PR interval was significantly longer in group A (N = 16) compared to group B (N = 41), 140 ± 24 ms vs. 121 ± 13 ms (p < 0.01). Six cases of 1°AVB (one with intermittent 2°AVB) and one case of intermittent 1°AVB developed in group A, progressing from normal sinus rhythm at 1 month of age. Prenatal Doppler predicted development of 1°AVB at follow-up with a sensitivity of 100%, a positive predictive value of 37.5%, a likelihood ratio of +5.1 and a negative predictive value of 100%. No differences were revealed regarding HR, QTc, or M-mode measurements. Children in group A had a slightly higher myocardial performance index, mainly due to a longer isovolumetric contraction time, in both flow- and tissue Doppler imaging (TDI) recordings. Abnormal TDI values were seen in only one case. **Conclusions:** Fetally anti-SSA/Ro exposed children with transient perinatal signs of disturbed AV conduction may progress to 1°AVB postnatally. Children at risk can be identified by prenatal Doppler echocardiography.

#### P-27

##### **Analysis of QT dispersion and corrected QT interval in children with hypertrophic cardiomyopathy**

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**Introduction:** Hypertrophic Cardiomyopathy (HCM) in children and adolescents is associated with an increased risk of ventricular arrhythmias which may originate from repolarization disturbances. Study aimed to assess QT dispersion (QTd) and corrected QT interval (QTc) in children with HCM.

**Material and Methods:** Study included 32 children with HCM (4–18 years; 14 girls) and 58 healthy children (4–19 years; 25 girls). In all children the physical examination was carried out, as well as echocardiographic examination and resting ECG



tracing (50 mm/s) with the assessment of QTc and QTd. By means of Mann-Whitney test the continuous values of QTc and QTd were compared between analyzed groups. Continuous variables were presented as median and IQR. Fishers exact test was used to analyze the rate of prolonged QTd values  $\geq 50$  ms and QTc  $\geq 440$  ms. The logistic regression analysis was carried out to estimate Odds ratio (OR) with 95% confidence interval (CI) for occurrence of the abnormal QTd and QTc values, depending on the presence of HCM.

**Results:** In children with HCM, in comparison with the control group, there were noted significantly higher values of QTd: 40 (30–50) ms vs. 20 (20–30) ms ( $p < 0.0001$ ) and QTc: 423 (397–446) ms vs. 391 (386–401) ms ( $p < 0.0001$ ).

The values QTd  $\geq 50$  ms ( $p < 0.01$ ) and QTc  $\geq 440$  ms ( $p < 0.0001$ ) occurred significantly more frequently in group of children with HCM. QTd  $\geq 50$  ms was identified in 6 (19%) children with HCM. In 7 (22%) children with HCM the QTc value exceeded 440 ms. No abnormal values of QTd or QTc were noted in any child from the control group. The presence of HCM was related to increased risk of the occurrence of QTd  $\geq 50$  ms (OR 3.7; 95% CI 19.7–5277.6;  $p < 0.0001$ ) and QTc  $\geq 440$  ms (OR 5.7; 95% CI 20.2–5277.6;  $p < 0.0001$ ).

**Conclusions:** The presence of HCM in children is associated with the increase of dispersion and with the prolongation of QT interval.

#### P-28

##### **Analysis of the signal-averaged QRS complex width in healthy children and in children with hypertrophic cardiomyopathy**

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**Introduction:** Hypertrophic cardiomyopathy (HCM) is associated with ventricular arrhythmias and sudden death. Signal-averaged ECG (SAECG) analysis is used to detect electrical signals of small amplitude that are not otherwise evident. The aim of the study was to assess the SAQRS in healthy children and in children with HCM.

**Material and methods:** The study included 32 children with HCM (4–18 years; 14 girls) and 58 healthy children (4–19 years; 25 girls). In all patients physical examination was carried out, as well as echocardiographic examination and SAECG with the usage of high-pass filter settings with cut-off frequencies 25–250 and 40–250 Hz. Statistical analysis was carried out by means of Mann-Whitney test, Fisher's exact test and non-parametrical Spearman's rank correlation. Continuous variables were presented as median and IQR. A probability of  $< 0.05$  was considered statistically significant.

**Results:** Late ventricular potentials (LP) occurred more frequently in children with HCM in comparison with control group (22% vs. 7%,  $p < 0.05$ ). The presence of HCM was associated with more frequent occurrence of LP (21.9% vs. 6.9%;  $p = 0.049$ ). In comparison with healthy children the SAQRS duration was significantly longer in children with HCM [25–250 Hz: 117.0 (102.0–124.5) ms vs. 92.0 (86.0–100.3) ms,  $p < 0.0001$ ; 40–250 Hz: 114.0 (99.3–121.8) ms vs. 92.0 (85.8–98.3) ms,  $p < 0.001$ ]. The duration of SAQRS  $\geq 114$  ms occurred significantly more frequently in children with HCM (25–250 Hz: 53% vs. 0%,  $p < 0.0001$ ; 40–250 Hz: 47% vs. 0%,  $p < 0.0001$ ).

In the group of children with HCM significant correlations were observed between SAQRS duration (filters cut-off frequencies 25–250 Hz) and ventricular septum thickness (25–250 Hz:  $r = 0.44$ ,  $p = 0.012$ ; 40–250 Hz: ( $r = 0.44$ ;  $p = 0.013$ ) and left ventricular mass index (25–250 Hz:  $r = 0.52$ ,  $p = 0.002$ ; 40–250 Hz:  $r = 0.49$ ;  $p = 0.005$ ).

**Conclusions:** Cardiac remodeling observed in children with HCM is associated with more frequent occurrence of LP and increased duration of ventricular depolarization. Duration of ventricular depolarization in children with HCM depends on the ventricular septum thickness and left ventricular mass.

#### P-29

##### **Vectorcardiographic measurements of QT interval in a paediatric LQTS population**

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**Introduction:** Due to the high and variable heart rate in a paediatric population, measurements of corrected QT interval for heart rate (QTc) are even less reliable than in adults. Computerized electrocardiograms are now widely used but it has been shown that the diagnostic interpretation provided by the automated analysis system may fail to identify many carriers of mutations causing LQTS.

The aim of this study was to investigate if vectorcardiogram (VCG) according Frank lead system could be superior to 12-lead ECG in providing correctly LQTS diagnosis in children.

**Material and Methods:** The LQTS population consisted of 35 genetically confirmed carriers of mutations in the KCNQ1 gene ( $n = 29$ ) and KCNH2 gene ( $n = 6$ ). The control group comprised of 35 age and gender matched healthy children (of which 10 were confirmed non-carriers). Mean age in the LQTS group and the control group was 7.0 and 6.7 years respectively (range 0.5 to 16 y). There were 20 girls and 15 boys in each group.

Standard 12-lead ECG and VCG were recorded in all study participants. QT intervals were measured manually by one author (AW) blinded to the patients genotype and with a documented small intraobserver relative error (1.3%). The 12-lead ECG automatic measurements and interpreting of QTc, were performed with a Mac<sup>®</sup>5000 (GE Medical system) and the VCG automatic measurements were made with Mida<sup>®</sup>1000, CoroNet (Ortivus AB). A QTc  $> 440$  ms by either method was considered prolonged and indicative of LQTS.

**Results:** 30 LQTS children out of 35 (83%) were correctly diagnosed using the VCG automatic measurements of the QTc. The manually assessed QTc, automatic measured QTc (ECG) and automatic interpreting from standard ECG correctly diagnosed 29 (82%), 24 (69%), 17 (49%) respectively, of 35 LQTS children.

**Conclusions:** This study showed comparable results for VCG automatic measurements and a highly experienced observer in the ability to provide correct LQTS diagnosis in children, based on QTc measurements. The automatic interpreting of the ECG showed a poor ability to render correct diagnosis in a paediatric LQTS population.

#### P-30

##### **First Results from the Austrian LQTS Registry**

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**Aim:** The registry was initiated in 2006 as a multicentre project aiming at collecting data from all patients with documented LQTS up to the age of 18 years. Approval was given by the ethics committee. **Results:** By the end of 2010 the registry contains data from 47 patients (23m, 24f) with an actual age from 3 to 22 years. 26 patients (14m, 12 f)(55%) presented with symptoms. Mean age at diagnosis was 3.5 years in male and 6.5 years in female patients. Symptoms were stress associated syncope, palpitations and documented ventricular tachycardia (TdP). 17 patients (36%) are still asymptomatic. 4 patients (3 m; 9%) died suddenly. By the end of 2010 all 43 surviving patients are treated with  $\beta$ -blockers, 10 had either an implantation of a pacemaker (4), or an ICD (6). Genetic analyses were performed in 38 patients and revealed presence of LQT 1 in 17(6m, 11f), LQT 2 in 8 (3m, 5f) and LQT 3 in one patient (m). The remaining results are still pending. In addition to collecting data from patients diagnosed in the participating paediatric cardiology units it was possible to identify 39 family members with so far undiagnosed LQTS (16m, 23f). From 2006 to 2010 we noted a relative increase of newly diagnosed patients.

**Summary:** The registry is a useful instrument for providing data both on actual numbers and treatment of patients with LQTS less than 18 years in our country. Annual data presentation in the working group leads to more awareness of LQTS and may also contribute to the reduction of secondary deaths in families.

### P-31

#### **Tilt-table and ergometry tests in relation to cardiac status**

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Ergometry and tilt-table test are non-invasive diagnostic tools for cardiac status evaluation of patients with arrhythmia, chest pain or syncope, whose negative findings exclude cardiac causes.

The aim of this study was to evaluate these methods in paediatric patients. The investigation was performed at Clinical Centre of University of Sarajevo, Paediatric Clinic: Department of Cardiology, during April 2008 till April 2010. Step ergometry was performed in all pts with continuous monitoring of: blood pressure, electrocardiogram, oxygen saturation and symptoms, according to Naughton protocol 2.0. A total number of 119 patients were tested. Group of 75 patients age 7–18 years (mean age 13.4 years) was subjected to the ergometry: 41 boys (54.7%) and 34 girls (45.4%). The most common diagnoses of evaluated patients by ergometry were: arrhythmio cordis 27 (36%), stenocardiae 32 (42.7%), hypertensio arterialis 6 (8%) and others. A positive finding was diagnosed in 17 patients (22.6%) and negative in 58 patients.

Tilt-table test was performed in 44 patients: age from 8 years till 18 years (mean age 13.9 years), 19 boys (43.2%) and 25 (56.8%) girls. The majority of patients observed by tilt-table test had syncope in 41 (93.2%), vertigo 2 patients (4.5%) and others. Positive findings were detected in 27 patients (61.4%) and negative findings in 17 patients (38.6%). These results were helpful in establishing diagnosis of syncope aetiology as the most common cause for performing this test. However, ergometry was

negative even in 58 patients, excluding the cardiac cause of disease.

**Conclusion:** These results are the first experience of Sarajevo school in performing these tests. Ergometry and tilt-table test are valid non-invasive diagnostic tools in assessing cardiac status of paediatric patients.

### P-32

#### **Changes of Gene Expressions in Monocrotaline induced Pulmonary Hypertension Rats after Bone Marrow Cell Transfusion**

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Pulmonary artery hypertension (PAH) causes right ventricular failure and possibly even death by a progressive increase in pulmonary vascular resistance. With a progressive loss of pulmonary microvasculature, it later becomes refractory to traditional therapies. Bone marrow-derived mesenchymal stem cell therapy has provided an alternative for ailments of various organs by regeneration at the site of a lesion. The purposes of this study were to investigate changes of pulmonary pathology and investigate changes of gene expressions of ET (endothelin)-1, ET receptor A (ERA), endothelial nitric oxide synthase (eNOS), matrix metalloproteinase (MMP)2, tissue inhibitor of matrix metalloproteinase (TIMP), interleukin (IL)-6, tumor necrosis factor (TNF)  $\alpha$  in monocrotaline (MCT)-induced pulmonary hypertension rat models after bone marrow cell infusion.

**Methods:** The rats were grouped as follows: control group, subcutaneous (sc) injection of saline; MCT group, sc injection of MCT; bone marrow cell infusion (BM) group, sc injection of MCT plus bone marrow cell infusion by intravenous injection at the tail 1 week after MCT injection.

**Results:** The average RV pressure in week 4 significantly increased in the MCT group compared with control group. It had significantly decreased in the BM group compared with the MCT group. RV weight had significantly decreased in week 4 in the BM group. The ratio of right heart/left heart+septum had significantly decreased in weeks 3 and 4 in the BM group compared with the MCT group. Medial wall thickness of pulmonary arterioles and number of muscular pulmonary arterioles had minimally decreased in the BM group compared with the MCT group. However, there was not any significant difference between the two groups. Gene expressions of ET-1, ERA, eNOS, MMP2, TIMP, IL-6 and TNF  $\alpha$  had significantly increased in the MCT group after week 3 and significantly decreased in week 4 in the BM group compared with the MCT group.

**Conclusion:** There was improvement of RVH and mean RV pressure after bone marrow cell infusion. Decreases in several gene expressions were observed. Additional research on the dose and frequency of bone marrow infusion is needed to better determine the appropriate amount of bone marrow cell count required for PAH treatment.

### P-33

#### **Apoptosis associated Gene Expressions of in Monocrotaline-Induced Pulmonary Hypertension in Rats after Bosentan Treatment**

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**Purposes:** Vascular wall remodeling in pulmonary hypertension is contributed to by an aberration in the normal balance between proliferation and apoptosis of smooth muscle. Endothelin-1 prevents up-regulation of pro-apoptosis and activates caspase-3. Endothelin (ET)-1, a potent endothelium-derived vasoconstrictor peptide, has several properties suggestive of its potential pathophysiological role in pulmonary hypertension.

The objectives of this study were to evaluate the effect of bosentan on bcl<sub>2</sub>, caspase-3, vascular endothelial growth factor (VEGF), interleukin(IL) 6 and tumor necrosis factor (TNF)-α in monocrotaline (MCT)-induced pulmonary hypertension and to investigate the correlation between caspase-3 gene expression and other associated genes.

**Methods:** Sprague-Dawley rats were divided into three groups: control group, MCT group (60 mg/kg), bosentan group (MCT 60 mg/kg plus 20 mg/day bosentan orally). The animals were sacrificed after 1 day, 5 day, 1 week, 2 week and 4 week. Gene expressions of bcl<sub>2</sub>, caspase 3, VEGF, IL-6 and TNF-α were analyzed by real time polymerase chain reaction and western blot analysis.

**Results:** The gene expressions of bcl<sub>2</sub>, caspase 3, VEGF had significantly increased in the MCT group compared with the control group in week 4 by PCR analysis. The gene expressions of caspase and VEGF had significantly decreased in the bosentan group compared with the MCT group in week 4. Bcl<sub>2</sub> protein content had significantly increased in the bosentan group in week 4. Gene expressions of IL-6 had significantly increased in week 1, 2 and 4 in the MCT group compared with the control group. Gene expressions of IL-6 had significantly decreased in week 1, 2 and 4 in the bosentan group compared with the MCT group. Gene expressions of TNF-α had significantly increased on day 5 in the MCT compared with the control group. Gene expressions of TNF-α had significantly decreased on day 5, in week 1 and 2 in the bosentan group compared with the MCT group. Caspase-3 gene expression significantly correlated with bcl<sub>2</sub> and VEGF gene expressions.

**Conclusion:** Bosentan may hold considerable potential for preventing apoptosis and inflammation.

#### P-34

##### Rapidly activating delayed rectifier K<sup>+</sup> current during post-natal development in mouse ventricle

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**Introduction:** The rapidly activating delayed rectifier K<sup>+</sup> current (IKr) plays an important role in the repolarization of cardiac action potentials in many mammalian species and is encoded by the ether-a-go-go-related gene (ERG). There is some evidence that IKr is developmentally regulated in mouse heart. However, in these studies, neonatal mouse ventricular myocytes were isolated using the 'chunk' method, which was previously demonstrated to damage the delayed rectifier K<sup>+</sup> current. The present study aimed at investigating the developmental changes in IKr in mouse ventricular myocytes isolated from the same Langendorff perfusion method.

**Methods:** Single ventricular myocytes were enzymatically isolated from various post-natal stages (day-0 to adult) of C57BL/6 mice using the same Langendorff perfusion method and stored in normal Tyrode solution containing 1.8 mM Ca<sup>2+</sup>. Appearance of viable day-0 neonate ventricular myocytes was rather rod-shaped, similar to that of adult ventricular myocytes. Whole-cell

patch-clamp method was used to record IKr. IKr was identified as an E-4031-sensitive current during various test potentials applied from a holding potential of -50 mV and was expressed as current density (pA/pF). The expression of ERG proteins was also quantitatively assessed using Western blotting.

**Results:** IKr was the dominant outward K<sup>+</sup> current in day-0 neonatal ventricular myocytes and its amplitude was gradually decreased during the development; IKr amplitude in day-0 and adult myocytes were 3.3 ± 0.2 and 0.5 ± 0.1 pA/pF, respectively. The extent of prolongation of action potential duration at 90% repolarization (APD<sub>90</sub>) by E-4031 was also progressively decreased from day-0 (30 ± 6%) and day 7 (25 ± 8%) neonate to adult ventricular myocytes with negligibly small effect. Western blot analysis revealed that ERG protein expression detected at the levels of 130 and 155 kDa was also gradually decreased during development from neonate to adult.

**Conclusions:** These data demonstrate that expression of IKr channel and its functional role is developmentally regulated in mouse ventricular myocytes.

#### P-35

##### Postnatal developmental changes in the TRPC channels in mouse cardiomyocytes

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**Introduction:** Evidence has been presented to indicate that there are postnatal developmental changes in cardiac Ca<sup>2+</sup> channels and transporters, such as sarcolemmal L-type channels, Na<sup>+</sup>/Ca<sup>2+</sup> exchangers, and the sarcoplasmic/endoplasmic reticulum Ca<sup>2+</sup> ATPase (SERCA2a). The transient receptor potential canonical (TRPC) channels are Ca<sup>2+</sup>-permeable nonselective cation channels widely expressed in diverse cell types including cardiac myocytes and are typically activated following depletion of endoplasmic/sarcoplasmic reticulum (ER/SR) Ca<sup>2+</sup> stores. The present study was undertaken to investigate the possible developmental changes in TRPC channels in mouse ventricles.

**Methods:** Single ventricular myocytes were enzymatically isolated from various postnatal stages (day-0 to adult) of C57BL/6J mice using a retrograde Langendorff perfusion method. Membrane currents were measured using whole-cell patch-clamp methods with Cs<sup>+</sup>-rich pipettes and K<sup>+</sup>-free bath solution and were expressed as current density (pA/pF). The expression of TRPC isoforms was quantitatively assessed by Western blotting.

**Results:** Bath application of thapsigargin (2 μM) gradually activated the membrane current that exhibited a practically linear I-V relationship with a reversal potential of ~0 mV, a sensitivity to inhibition by 2-aminoethoxydiphenyl borate (20 μM, 2-APB) and marked reduction in inward current after replacement of extracellular Na<sup>+</sup> with N-methyl-D-glucamine (NMDG). Based on these properties, the thapsigargin-induced current was identified as the TRPC current. The amplitude of TRPC current measured at +40 mV peaked in day-0 neonatal ventricular myocytes (5.8 ± 1.2 pA/pF) and declined thereafter during postnatal development to 1.8 ± 0.9 pA/pF in adult ventricular myocytes. Western blot analysis revealed that expression of TRPC1, 3, 4, 5 proteins was also gradually decreased during development from neonate to adult.

**Conclusions:** Our results provide experimental evidence to show that expression of TRPC channel is developmentally regulated in the mouse ventricle with a higher level in neonatal compared with adult ventricular myocytes.



## P-36

**Right ventricle function analysis by using RV papillary muscle**

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**Introduction:** Right ventricular dysfunction is a common long-term complication in patients after the repair of congenital heart disease. Previous investigators have examined the cellular and molecular mechanisms of left ventricular (LV) remodeling, but little is known about the stressed RV. Our purpose was to provide a detailed physiological characterization of a model of RV hypertrophy.

**Method:** A right lateral thoracotomy was performed on SD rat at the weight of 200 g. Then, the main pulmonary artery was banded with a 5-0 suture, tied tight against a 25G needle. RV pressure was examined by using 1.2F micro cath. at 4 weeks postoperative. The rats with elevated RV pressure ( $>80$  mmHg) were enrolled for more further study. Age matched SD rats were examined for control. We used aequorin method to evaluate tension development with  $Ca^{2+}$  in right ventricular papillary muscle in place of RV function.

**Results:** RV pressure was significantly difference between PAB and control ( $88.2 \pm 13.9$  mmHg vs.  $21.8 \pm 5.1$  mmHg,  $P < 0.001$ ). There was no significant difference in cardiac output ( $0.38 \pm 0.09$  vs.  $0.37 \pm 0.11$ ; NS). RV weight increased by 70% without significant change in LV weight. We confirmed that peak  $Ca^{2+}$  in PAB was significantly higher than that in control ( $2.35 \pm 0.03$  vs.  $1.52 \pm 0.11$   $\mu$ M,  $p < 0.05$ ). Interestingly, peak tension of both cardiac muscle was not significantly different at 4 weeks after operation ( $26.52 \pm 9.91$  vs.  $38.03 \pm 5.97$  mN/mm<sup>2</sup>). We also analyzed time courses in tension and  $Ca^{2+}$ . Relaxation time in PAB was not significantly different from that in control ( $64.00 \pm 2.89$  vs.  $70.67 \pm 2.82$  msec), however, decay time of light in PAB was significantly slower than that in control ( $45.33 \pm 3.76$ ,  $27.33 \pm 1.15$  msec,  $p < 0.05$ ).

**Conclusions:** RV hypertrophy could have impairment of  $Ca^{2+}$  handling possibly due to impairment of  $Ca^{2+}$  handling. This model, and the method are useful to understand the RV dysfunction. More further study will be needed to establish the strategy for RV failure.

## P-37

**Infective Endocarditis in patient with congenital heart disease and without vegetations**

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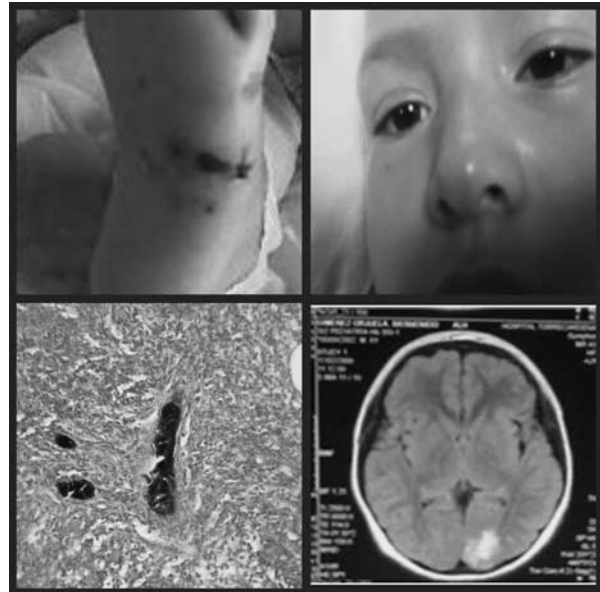
**Introduction:** Infective endocarditis is a fatal disease without treatment. Early diagnosis and therapeutic approach are very important to reduce morbidity and mortality. Given the characteristics and pathogenesis of this disease, sometimes the diagnosis is complex and involves a wide differential diagnosis.

**Case report:** A 6 years old children is attended by pediatrics emergency with vomiting and fever of 24 hours duration. In the last hours began with pain and signs of inflammation in his left foot plant.

**Family History:** Colombian parents, without interest.

**Personal History:** Bicuspid aortic valve with severe stenosis who underwent surgery 6 years ago. Currently has a dual moderate aortic injury. Travel to Colombia which he returned for 16 days.

**Exploration:** Systolic murmur III/VI. Increased temperature and edema in his left ankle, signs of inflammation of subcutaneous tissue in the left foot plantar region with bruise damage (figure 1). Hiperesthetic greyish lesion on tip of nose. Inflammatory signs in 5th finger right hand. Diffuse abdominal pain without peritoneal irritation. Severe headache with no neurological deficit.



**Differential Diagnosis:** infective endocarditis, primary systemic vasculitis, infectious vasculitis.

**Investigations:** CBC: leukocytosis with neutrophilia. Increased acute phase reactants. Rx: cardiomegaly, no condensation in lung parenchyma. Echocardiography showed vegetations or not changes in basic valve disease. ANCA negative. Negative immune. Normal complement. Cryoglobulins negative. Serology Leptospira, Rickettsia, Borrelia Dengue and negative. Blood cultures (2) positive *S. aureus methicillin sensitive*. Biopsy of skin lesions, septic emboli source (figure 2).

**Evolution:** On admission he was treated for infective endocarditis antibiotic (ceftriaxone, vancomycin, gentamicin) to receive blood cultures (Cloxacillin).

According to the Duke criteria we support the diagnosis of bacterial endocarditis. As a complication he had septic emboli brain (figure 3, MRI). The outcome was favorable, the patient was asymptomatic at present.

**Conclusion:** Infective endocarditis is a rare disease in childhood, although their frequency is increasing due to longer survival of children with congenital heart disease. The diagnosis can be used DUKE criteria, taking into account that the absence of vegetation does not exclude the diagnosis.

## P-38

**Factors influencing on formation of heart rhythm disturbances in newborns**

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**Objective:** To determine risk criteria of formation and advance of heart rhythm disturbances in newborns.

**Methods:** Data of gynecologic and obstetric history, ECG, 24-hour monitoring with assessment of rhythm variability, Echo, neurosonography, thyroid hormones, markers of myocardial damage and antibodies to myocardium tissue tests are used.

**Results:** 102 patients (14 healthy incl.) were examined. Extrasystoles had the most specific gravity in the structure of idiopathic arrhythmias in newborns – 32,4%. Bradyarrhythmias were 25,7%, tachyarrhythmias – 22,9%, WPW syndrome – 18,9%. Rhythm disturbances preserve only in 5,4% by the sixth month of life. Longer persistence is typical for extrasystole and WPW syndrome.

Heart rhythm disturbances are marked much more often in newborns whose mothers had acute respiratory disease during pregnancy ( $p = 0,049$ ), and who were born from the primipregnancy ( $p = 0,041$ ). Bradyarrhythmias and tachyarrhythmias have similar factors which can potentially favor arrhythmia manifestation: intracranial hypertension according to neurosonography and changes of hormonal profile of thyroid body towards hypofunction. Established fact that higher value of systolic pressure in the right ventricle (average 28,38 mmHg ( $p = 0,047$ )) is the peculiarity of intracardiac hemodynamics in the group of newborns with heart rhythm disturbances.

Presence of extrasystole in newborns is related with increase of troponin level I ( $p = 0,015$ ) and activation of parasympathetic link of vegetative nervous system (increase of pNN50 ( $p = 0,009$ ) and SDNNi ( $p = 0,037$ ) in comparison with healthy children). Association of bradyarrhythmias with level of myocardial anti-nuclear antibodies in blood ( $\chi^2 = 4,89$ ;  $p = 0,027$ ) are marked for fact in newborns. This fact says for significance of immune factor in damage of myocardium and its conduction system.

**Conclusion:** Thus, autoimmune component is important link of bradyarrhythmias pathogenesis, which, probably, is formed into antenatal period with the help of maternal antibodies. Destructive processes in myocardium, accompanying by increase of troponine I level in blood serum, and also activation of parasympathetic link of vegetative nervous system during extrasystole depend, on the contrary, on factors connecting with intra- and postnatal periods. Infection during pregnancy can influence, indirectly, on the process abnormality of obliteration of additional conduction tracts at WPW syndrome.

### P-39

#### RFA of tachyarrhythmias in children of one year old

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**Objective:** It is known that RFA is the only available treatment of small children with drug refractory tachyarrhythmias. The aim of our abstract is to show our experience of RFA of tachycardias in small children.

**Materials:** Twelve RFA of tachyarrhythmias were performed to children of 1–12 months old in our clinic during last five years. The minimum age of effective RFA is 48 days, the minimum weight is 3,8 kg. Tachycardia was first disclosed in three children antenatally. Tachycardia was incessant in 8 cases and paroxysmal in 4 cases. The presence of arrhythmogenic cardiomyopathy and also ineffectiveness of antiarrhythmics combinations were the indications for RFA performance in all cases.

**Results:** Five children had WPW syndrome. Localization of accessory pathway: left posterior ( $n = 2$ ), left anterolateral ( $n = 2$ ) and right posteroseptal ( $n = 1$ ). In all cases of left sided localization of accessory pathway an approach into the left atrium was carried out through the patent foramen oval. Seven children had intra-atrial tachycardia. Localization of atrial ectopic

focuses was determined in the area of right atrial auricle basis ( $n = 2$ ), in the area of right atrial anterior wall ( $n = 3$ ), in the area of His band ( $n = 1$ ), in the area of the patent foramen oval ( $n = 1$ ). Navigation mapping was performed to three children with intra-atrial tachycardia. The first RFA attempt was in two children at the age of one month, but the procedure was ineffective. Antiarrhythmic therapy had temporary effect, and at the age of 4–5 months RFA of accessory pathway and right atrial ectopic tachycardia was successfully performed. Intra- and postoperative complications were not observed. According to the Echo data reduction of atrium sizes, increase of left ventricle contractile function were marked in 5–10 days ( $p < 0,05$ ). Total RFA effectiveness in children till one year old is 100%. Follow-up was from one month to five years. Tachycardia relapses were not disclosed. According to Echo, pathology was not discovered. **Conclusion:** RFA is an effective and safe method of tachyarrhythmia treatment including infants. All children of early age with hemodynamic instability, drug refractory should be turned to specialized centers, having RFA experience at the given age.

### P-40

#### Properties of heart remodeling in children with congenital heart diseases complicated by pulmonary hypertension

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**Objective:** We aimed to assess echocardiography data of heart remodeling in children with congenital heart diseases (CHDs) and different degrees of pulmonary arterial hypertension (PAH).

**Methods:** 422 echocardiography protocols were analyzed. The Echo was performed to children from 1 month to 18 years. 284 (67,3%) were defined having PH 1 degree, 52 (12,3%) – PH 2 deg., 72 (17,1%) – PH 3 deg., 14 (3,3%) – PH 4 deg. PH degree was defined by the level of systolic pressure in the right ventricle (SPRV) according to the Echo: 30–50 mmHg – PH 1 deg., 51–69 mmHg – PH 2 deg., 70 mmHg and higher – PH 3–4 deg. Echo data were showed in percentage of individual norm. End diastolic volume of left ventricle (EDV LV), its form: spherical (SI) and eccentric index (EI), volume of right and left atrium, LV ejection fraction and contractility index of right ventricle, index of systolic (ISR) and index of diastolic remodeling (IDR) were assessed. Data are presented as  $M \pm SD$ , where M is an average value; SD is standard deviation.

**Results:** PH growth from 1 to 3 degree is accompanied by EDV LV increase to  $157,59 \pm 98,54\%$  ( $p < 0,001$ ), and at 4 degree of PH EDV decrease is observed to  $75,11 \pm 15,35\%$  ( $p < 0,001$ ). Significant change of LV form is marked at 4 degree of PH: SI is  $1,84 \pm 0,48$  r.u. and EI –  $0,55 \pm 0,49$  r.u. ( $p > 0,001$ ). Right atrium volume increase to  $132,37 \pm 24,15\%$  ( $p > 0,001$ ) is discovered at 2 degree of PH, left atrium volume increases to  $133,62 \pm 24,15\%$  ( $p < 0,001$ ) at 2 degree of PH, left atrium volume decrease to  $65,13 \pm 15,08\%$  ( $p < 0,001$ ) is observed at 4 degree of PH. Tendency of LV ejection fraction decrease as PH growths and reduction of right ventricle contractility index at PH of 4 degree to  $26,1 \pm 7,5$  r.u. ( $p < 0,001$ ) were determined. LV systolic remodeling was discovered in children with PH 2deg. ( $ISR-1,19 \pm 0,31$ r.u.), and diastolic remodeling at PH 3–4deg. ( $IDR-3,35 \pm 1,2$ r.u.) ( $p < 0,001$ ).

**Conclusion:** Changes of structural characteristics of heart chambers and their function disturbance are observed at CHDs with PH. LV remodeling with systolic dysfunction is appeared at CHDs complicated by PH of 2deg., and PH of 3–4 degree with LV diastolic dysfunction.

**P-41****Comparative assessment of intracardiac hemodynamics state in children with ventricular premature beats from RVOT and LVOT**

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**Objective:** To carry out comparative analysis of intracardiac hemodynamics state in children with ventricular premature beats (VPBs) from RVOT and LVOT, and also its condition after RFA of arrhythmias focus.

**Methods:** Patients of both sexes were included into the study. They had ventricular arrhythmia more than 20% of overall quantity of heart beats per day and without organic and structural heart pathology. The first group consisted of 13 pts at the age of 12–17 years old ( $13 \pm 3,7$  years) with extrasystole from RVOT. The second group involved 11 pts at the age of 13–16 years old ( $14 \pm 2,3$  years) with extrasystole from LVOT. The control group included healthy volunteers at the age of 12–17 years old ( $13 \pm 3,3$  years). Method of quantitative blood pool SPECT was applied for intracardiac hemodynamics assessment.

**Results:** Comparative hemodynamics analysis between the first and the control groups showed no significant differences. In comparison with the control group, decrease of LV EF was disclosed in the group of patients with extrasystole from LVOT. Significant differences of hemodynamics state between the first and the second groups were not discovered. After RFA significant EF increase either of LV or RV was disclosed, stroke volume of RV increased, peak filling rate and mean filling rate of RV for 1/3 diastole increased, time till peak filling rate of RV decreased.

**Conclusions:** Ventricular premature beats from RVOT result in diastolic dysfunction of right ventricle. VPDs from LVOT are accompanied diastolic dysfunction either of right or left ventricles. RFA of arrhythmias focus results in indices normalization of intracardiac hemodynamics.

**P-42****Cardiotrophin-1 is differentially induced in the myocardium of infants with congenital cardiac defects depending on hypoxemia**

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**Objective:** To test if cardiotrophin (CT)-1 is differentially induced in the myocardium of infants with congenital cardiac defects depending on hypoxemia.

**Background:** CT-1 is up regulated by hypoxemia and hemodynamic overload and induces cardiac hypertrophy via the janus kinase/signal transducer and activator of transcription pathway.

**Methods:** Infants with tetralogy of Fallot (TOF) or with large ventricular septal defect (VSD) undergoing corrective surgery were investigated. Expression of CT-1 was assessed at mRNA- and protein level in the right atrial and -ventricular myocardium. We measured the activation of the signal transducer and activator of transcription (STAT)-3, vascular endothelial growth factor (VEGF)165, phosphorylated extracellular regulated kinase (ERK)-1/2 MAP kinase and heat shock proteins (HSP)-70 and -90. Degradation of cardiac troponin (cTn)-I served as a marker of myocardial damage.

**Results:** CT-1 was detected in all patients. Infants with TOF showed significantly higher protein levels of CT-1 than those with VSD. In all patients, levels of CT-1 negatively correlated with arterial oxygen saturation. Patients with TOF showed higher levels of phospho-STAT-3 and VEGF165 and higher levels of cTn-I. In contrast, the activation of ERK1/2 MAP kinase and levels of HSPs were not different in both groups.

**Conclusions:** Our results show that CT-1 is differentially expressed in the myocardium of infants with heart defects. Higher CT-1 expression in patients with TOF is associated with activation of the Jak/STAT pathway and higher cTn-I degradation. CT-1 might mediate myocardial hypertrophy and myocardial dysfunction in infants with congenital cardiac defects, particularly in those with hypoxemia.

**P-43****Coronary ostia patterning in conotruncal defects**

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**Introduction:** Abnormal coronary ostia are frequently associated with conotruncal defects particularly with common arterial trunk. Our hypothesis is that these anomalies of coronary ostia in human could be related to variations in the location and size of the myocardial subpulmonary and subaortic domains.

**Methods:** In order to determine if abnormal outflow tract development influences coronary ostia position, we reviewed heart specimens with conotruncal defects: 46 common arterial trunk (CAT), 29 tetralogy of Fallot with pulmonary atresia (TOF&PA), 15 tetralogy of Fallot (TOF), 11 double-outlet right ventricle with subaortic ventricular septal defect (DORV) and 17 normal anatomy (control). Position of the coronary ostia over the aortic or truncal circumference was measured in degrees as the direction from the middle of the valvar orifice.

**Results:** The left coronary ostium was more posterior in conotruncal defects vs control (mean angle: control = 0°, TOF = 31°, TOF&PA = 47°, DORV = 44°, CAT = 65°,  $p < 0.005$ ) especially in CAT vs. other conotruncal defects ( $p < 0.05$ ). The right coronary ostium was more anterior in TOF, TOF&PA and DORV vs control (mean angle: control = 213°, TOF = 242°, TOF&PA = 245°, DORV = 271°,  $p < 0.05$ ) especially in DORV vs TOF and TOF&PA ( $p < 0.05$ ), but not in CAT (195°). The anterior intercoronary angle, which corresponds to the pulmonary identity domain, was similar in TOF, TOF&PA, DORV and control (133°–162°) but significantly larger in CAT (229°,  $p < 0.0001$ ).

**Conclusion:** Coronary artery ostia distribution is very disturbed in conotruncal defects. The type of abnormal pattern is different depending on the type of defect involved. In anomalies of rotation only (TOF, TOF&PA, DORV with subaortic VSD), the anterior intercoronary angle remains constant but the lateral shift of the coronary ostia varies according to the degree of outflow tract rotation. The marked difference between CAT and other conotruncal defects could reflect the impact of the absence of aortopulmonary septation on the width of the subpulmonary domain.

**P-44****Genetic polymorphism of methylenetetrahydrofolate reductase as a risk factor for congenital heart defect in Romanian children**

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**Introduction:** Congenital heart defects (CHD) are the most common single group of congenital abnormalities accounting for about 30% of the total abnormalities. Low folate intake as well as alterations in folate metabolism as a result of polymorphisms in the enzyme methylenetetrahydrofolate reductase (MTHFR) have been associated with an increased incidence of neural tube defects, vascular disease, congenital heart defects. To determine whether the C677T and G1793A variants in the MTHFR gene is associated with CHD susceptibility we utilised a case-control study.

**Methods:** DNA samples were isolated from peripheral blood samples and genotyped using polymerase chain reaction-restriction fragment length polymorphism (PCR-RFLP), with the digestion of restriction endonuclease *HinfI* and *BsrBI*. The restricted products were analyzed on 2% agarose gel.

**Results:** Subjects were consisted of 20 patients with CHD and 20 healthy controls. In patients, the genotypes frequencies of the MTHFR G1793A polymorphism were 22% and 88% for the GA and GG genotype respectively, whereas the genotypes frequencies of the MTHFR C677T polymorphism were 47.4%, 47.4% and 5.2% for CC, CT and TT genotypes respectively. The frequency of MTHFR 677CT+TT/1793GA was higher in the patients than controls.

**Conclusions:** The MTHFR C677T and G1793A polymorphism may influence congenital heart defect, but the MTHFR polymorphisms need to be studied further for confirmation in larger studies.

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#### P-45

##### **Interaction of Fetal Cardiology and Molecular Biology: About three cases of GATA4 deletion in fetuses with conotruncal malformations**

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We report three prenatally diagnosed cases of GATA4 deletion associated with conotruncal cardiac malformations. The first case is a fetus 20 weeks of gestation (Wks) with a complete and balanced atrioventricular canal. The second and third cases are fetuses aged 23 and 26 Wks with respectively a regular form of Tetralogy of Fallot (TOF) and a Truncus arteriosus type I. Classical and molecular cytogenetics studies on amniotic cells showed an interstitial deletion of the 8p23.1 region (RP11-23515 to RP11-589N15) encompassing the GATA4 gene locus, without modification of the telomeric region in the first two cases. The third case had been diagnosed in 1998, when molecular cytogenetic was not sufficiently developed to characterize the chromosomal rearrangement. Moreover, parents had refused further genetic studies. Conventional cytogenetic had, however found an *inv dup del 8p* with a GATA4 deletion. The first case was due to maternal transmission. The mother had the same deletion and she underwent surgery in childhood for a conotruncal malformation (interrupted aortic arch and malaligned ventricular septal defect) with good result. However, at 6 years of age, she presented severe mental retardation. Nobody in the family had the deletion. In the second case the parents decided to terminate pregnancy and the diagnosis of Truncus

arteriosus I was confirmed at autopsy. The third case TOF was repaired with no sequelae but the girl is very severely retarded. The chromosomal rearrangements in the second and third cases were *de novo*. The relevance of our cases, to our best knowledge, is the association between GATA4 deletion and conotruncal cardiac malformations, which is not been described in this pathology. Furthermore, we report the first case of maternal transmission (usually *de novo* or more rarely paternal transmission) of GATA4 deletion. We conclude that a multicenter study may be contributive to evaluate the prevalence of GATA4 deletion in conotruncal malformations diagnosed prenatally and therefore decide to perform or not on a routine basis the search for this deletion jointly with the deletion of 22.q11.1.

#### P-46

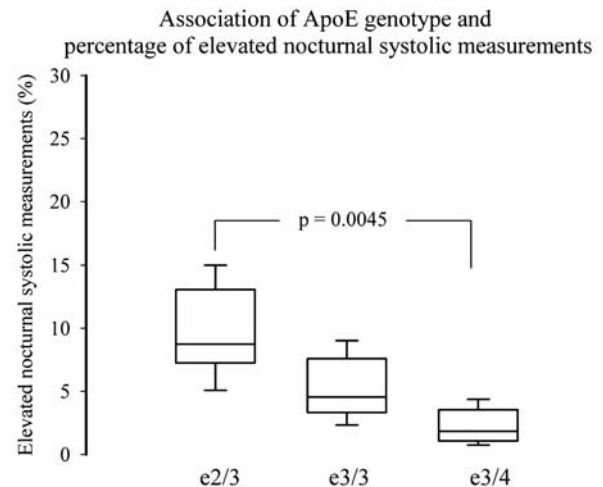
##### **Apolipoprotein e2 allele – a genetic risk factor for nocturnal blood pressure elevations in Type 1 diabetes**

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**Introduction:** We aimed to look for an association of the apoE genotype with blood pressure in adolescent patients with type 1 diabetes from Germany. Arterial hypertension, a precursor of cardiovascular disease in type 1 diabetes, is known to start as early as in childhood. Modified by gender and environmental factors, genetic variants of the apolipoprotein E (apoE) have been shown to influence the susceptibility to hypertension.

**Methods:** A total of 219 patients were recruited from the diabetes outpatient clinic. ApoE genotypes were determined by PCR and mass spectrometry analysis. Ambulatory blood pressure values were compared with the genotype.



**Results:** Patients with the e2/3 genotype compared to patients with the e3/4 genotype had higher nocturnal systolic blood pressure (mean sds 1.07 vs. 0.12,  $p = 0.022$ ). Moreover, patients with this genotype showed a higher percentage of elevated measurements of nocturnal systolic (7.26% vs. 1.07%,  $p = 0.0045$ ) and diastolic (4.74% vs. 1.54%,  $p = 0.017$ ) blood pressure. This association was confined to male and to non-obese patients.

**Conclusion:** The apoE e2/3 genotype is associated with elevated nocturnal blood pressure in a German male and non-obese population with type 1 diabetes. Apart from environmental

factors this genetic variant may increase susceptibility for cardiovascular morbidity and mortality.

#### P-47

##### **Coronary Flow and Reactivity, but not Arrhythmia Vulnerability, Are Affected by Cardioplegia During Cardiopulmonary Bypass Surgery of Piglets**

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**Introduction:** Cardiopulmonary bypass (CPB) surgery remains associated with significant cardiovascular morbidity in both paediatric and adult patients but the mechanisms are not fully clarified. Abnormalities in coronary flow and function have been suggested to play an important role. A few prior studies suggested protective effects on coronary and myocardial function by short intravenous (i.v.) infusion of cyclosporine A prior to CPB surgery.

**Methods:** Barrier-bred piglets (10–12 kg, n = 20) were subjected to CPB with (n = 10) or without (n = 10) antegrade administration for 20 minutes of cardioplegic solution. Prior to surgery, half of animals from each group received 10-minute i.v. infusion of 100 mg/kg cyclosporine A. Left anterior descending coronary flow velocity responses to adenosine, serotonin, and atrial pacing, as well as left ventricular function and postsurgical vulnerability to atrial fibrillation (Afib) were assessed by intracoronary Doppler, epicardial echocardiography, and in vivo electrophysiological study, respectively.

**Results:** Coronary peak flow velocity (cPFV) rose significantly after surgery, especially in cardioplegia group (p < 0.01 vs. non-cardioplegia group and pre-surgery). cPFV responses to adenosine, but not to serotonin, tended to decrease (p = 0.06) after surgery only in cardioplegia group (p = 0.06; p = 0.8 in non-cardioplegia group vs pre-surgery). After surgery, cPFV response to atrial pacing was lower in cardioplegia than in non-cardioplegia animals (p = 0.02). Neither vulnerability nor duration of induced Afib after surgery differed between groups (Chi-square p = 0.4). Cyclosporine had no significant effect on coronary indexes or arrhythmia vulnerability (p > 0.4). There was no difference in systolic myocardial function between groups at any timepoint.

**Conclusions:** Cardioplegia during CPB surgery of piglets was associated with profound abnormalities in coronary vasomotor tone and receptor-related flow regulation, whereas arrhythmia vulnerability appeared to be comparable with that in non-cardioplegia group. In this study, intracoronary pre-treatment with cyclosporine had no observable protective effect on coronary circulation or arrhythmia vulnerability after CPB surgery.

#### P-48

##### **The influence of right ventricular pressure overload on the expression of pro- and anti-inflammatory cytokines, growth factors and markers of apoptosis in the myocardium of newborn lambs**

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**Objective:** To assess the role of inflammatory mediators, growth factors and apoptosis on myocardial remodeling in a lamb model of moderate pressure overload of the right ventricle (RV).

**Methods:** 13 newborn lambs (age:  $3.3 \pm 1.3$  days) undergo either pulmonary arterial banding (PAB) (n = 6) or were sham operated (n = 7). Hemodynamic studies and blood samples taken for determination of cytokine levels were performed prior to and 1, 4 and 12 weeks after surgery. 12 weeks after the operation, RV biopsies were taken to analyse the intramyocardial expression of inflammatory mediators, growth factors and markers of apoptosis. Animals were then euthanised, hearts studied by morphology, histology and electron microscopy.

**Results:** In PAB animals a continuous increase of RV Pressure was registered (PA-RV pressure gradient after 12 weeks:  $56.67 \pm 8.43$  mmHg vs.  $5.43 \pm 1.51$  mmHg in controls, p < 0.001). This was associated with a significant hypertrophy of the RV as well as the LV myocardium. The RTQ-PCR analysis of the biopsies showed a slightly increased intramyocardial expression of all cytokines (TNF- $\alpha$ , IL-1 $\beta$ , IL-6, IL-8, IL-10), growth factors (CT-1, TGF- $\beta$ , VEGF) and markers of apoptosis (Bcl-XI, Bak, Fas-I) in PAB lambs vs controls. PAB group showed a significant increase in the intramyocardial expression of CT-1 as well as a systemic increase of IL-1 $\beta$ , IL-10, TGF- $\beta$  and Bcl-XI.

**Conclusion:** Our results show a measurable effect of moderate RV pressure overload on the systemic and intramyocardial expression of pro- and anti-inflammatory cytokines and an even more distinctive effect on growth factors and markers of apoptosis. The connected risk of myocardial remodeling and RV dysfunction should be expected. Systemic liberation of inflammatory mediators and growth factors might be responsible for LV hypertrophy observed in this model.

#### P-49

##### **Children with atrial septal defect and significant left-to-right shunt show myocardial remodeling**

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**Background:** Myocardial remodelling characterises a state of cellular and organ adaptation that involves inflammation, cell growth and cell death. The balance between mechanisms inducing cell growth and those inducing cell death by apoptosis determines whether cardiac function remains compensate or deteriorates in cardiac failure.

**Objectives:** To assess whether myocardial remodeling occurs in patients with atrial septal defect (ASD) with left-to-right shunt.

**Methods:** 15 children with ASD (10 females, age range 4–7 years, median 6.5 years) who were scheduled for surgical ASD closure were included. Myocardial specimens of the right atrium were obtained before connecting cardiopulmonary bypass. Gene expression of factors indicating mechanical stress (ANF), protective mechanisms (c-Fos, Hsp70, Hsp90), growth and angiogenesis (CT-1, HIF-1 $\alpha$ , VEGF), fibrosis of the extracellular matrix (PIIP), and regulating apoptosis (FasL, Bak, Bcl-xL) were determined by quantitative real time PCR.

**Results:** In all patients, expression of ANF, c-Fos, Hsp70, Hsp90 confirmed myocardial stress whereas expression of CT-1, HIF-1 $\alpha$ , VEGF and PIIP confirmed remodelling with signals

leading to cell growth, angiogenesis and fibrosis. All patients showed expression of FasL, Bak, Bcl-xL indicating apoptosis regulating mechanisms with a shift towards the expression of the apoptosis inhibitory factor Bcl-xL. Expression of factors inducing growth and angiogenesis correlated with the expression of factors responding to stress and also with the expression of apoptosis regulating proteins. Interestingly, expression of the apoptosis regulating proteins correlated with the marker of fibrosis PIIP.

**Conclusions:** Myocardial remodeling occurs in young children with ASD as a sign of compensatory adaptation and goes along with signals inducing hypertrophy, angiogenesis, fibrosis and apoptosis. In this stage of cardiac disease, antiapoptotic factors outbalance proapoptotic factors. A shift towards a net proapoptotic effect, which was not yet observed in this group, might mark the beginning of the transition from compensated to decompensated heart failure.

#### P-50

##### **A co-culture model that simulates Cardiopulmonary Bypass induced systemic inflammation: From bench to bedside**

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**Background:** Cardiopulmonary bypass (CPB) is known to induce a postoperative inflammatory response that is influenced by various factors. However, the underlying mechanisms are complex and not fully understood. To get more insights into the cellular mechanisms we developed an in vitro co-culture model that reflects the clinical situation. For evaluation of this model we compared the effect of hypothermia on the inflammatory response in the co-culture model with a clinical prospective study.

**Methods:** The co-culture model consisted of endothelial cells (HUVEC) and monocytes (THP-1). The cells were stimulated with 500 U/mL TNF-alpha to simulate CPB in pediatric open-heart surgery and exposed to moderate hypothermia (20°C) or normothermia (37°C). During the experiment, cell morphology (fluorescence microscopy), cell vitality (MTT-test) and cytokine release of IL-6 and IL-8 (ELISA) were investigated. In the prospective clinical trial, 20 patients (median age 3.3 months) undergoing CPB for ventricular septum defect (VSD) were randomized to receive either normothermic (37°C) or mild hypothermic (32°C) CPB. Dry blood spots (DBS) were obtained preoperatively, directly after weaning from CPB and after 24 h. IL-6 and IL-8 cytokines levels of the DBS samples were analysed by multiplexed sandwich immunoassays.

**Results:** We observed a significant IL-6 and IL-8 release in the co-culture model 2h and 24h after TNF- $\alpha$  stimulation. Clinically, the cytokine release was also seen directly after weaning from CPB and remained elevated until 24h. Interestingly, the IL-6 secretion in the co-culture 2h after TNF-alpha stimulation was significantly decreased under hypothermia. After 24h the IL-6 and IL-8 release of the co-culture model and the clinical data were similar and temperature independent.

**Conclusions:** These results demonstrate that our co-culture model is compatible to the clinical setting of pediatric CPB during VSD closure. The cytokine increase starts about the same time and is temperature independent after 24h in both, the in vitro co-culture and the prospective clinical trial. This may suggest that our co-culture model could be used for further studies on the mechanisms of CPB induced inflammation.

#### P-51

##### **Is mild hypothermia cool enough to protect cardiac cells after hypoxia? How does propofol affect this effect?**

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**Objectives:** The aim of this study was to elucidate cellular mechanisms responsible for the hypothermia induced protection of cardiac cells from apoptosis after a hypoxic event as observed cardiac arrest. Hence we investigated the role of therapeutic mild hypothermia (34°C) and propofol on cardiac cells after a hypoxic event.

**Methods:** Acute hypoxic conditions were simulated in a cardiac cell line (H9c2) using CoCl<sub>2</sub> [30 mM] for 1 h followed by a treatment with mild hypothermia and/or propofol [50  $\mu$ M] for 24 h. Cell survival was determined after 24 h of treatment using trypan blue staining. Apoptotic cellular activity was analysed by immunohistochemistry (M30-CytoDEATH) for cleaved keratin 18 as a caspase substrate. Regulation of key effector protein caspase 3 in the intrinsic apoptosis was investigated using western blotting and quantified by densitometry measurements.

**Results:** Mild hypothermic treatment (34°C) after hypoxia significantly increased survival of the cardiac cells after 24 h. Propofol treatment had no influence on cell survival after hypoxia. The combined application of mild hypothermia and propofol to cardiac cells did not show a significantly higher cell survival than treatment with mild hypothermia alone. Caspase triggered cleavage of keratin 18 occurred earlier and more extensive in normothermic cells. In densitometry analyses effector caspase 3 significantly decreased in cardiac cells starting at 6 h and continuing after 24 h of mild hypothermia.

**Conclusions:** Mild hypothermia is efficient enough to protect cardiac cells from apoptosis after a hypoxic event. Both, apoptosis process and execution is effectively reduced by mild hypothermia. Propofol treatment at 50  $\mu$ M after a hypoxic event does not have a significant influence on apoptosis in hypoxic cardiac cells, neither as singular nor as combined treatment with mild hypothermia.

#### P-52

##### **Modified Blalock-Taussig-Shunts: Histopathology and Morphometry**

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**Objective:** Surgical implantation of modified Blalock-Taussig-Shunts (mBTS) is a standard procedure to maintain pulmonary blood supply in cyanotic congenital heart defects. We are presenting results from histopathological work-up of a series of human explanted mBTS made of Gore-Tex fabric.

**Materials and methods:** Explanted mBTS (n = 9) were processed using a uniform protocol after surgical removal from pediatric patients with congenital heart disease (age at implantation 2 days to 4 months; time interval between implantation and explantation 4 months to 5 years). The specimen were fixed in formalin and embedded in paraffin or in methylmethacrylate. We performed standard staining, immunohistochemistry, and morphometrical analysis for quantification of pseudointima formation.

**Results:** On gross examination, mild to moderate proliferation of pseudointima was seen within the lumen of the mBTS



in 7 of 9 patients. Two of the shunts showed marked focal narrowing. The degree of pseudointima formation correlated positively with the implantation time. Superficial cells stained positive for von Willebrand factor (vWF) thus demonstrating complete endothelialisation. Neo-tissue formed within the mBTS contained fibromuscular cells (positive staining for smooth muscle actin, myosin, vimentin inter alia) as well as connective tissue. Some foreign body giant cells were seen locally related to the Gore-Tex material as well as few CD-68 positive inflammatory cells.

**Conclusion:** Histopathological work-up of mBTS revealed complete endothelialisation and only mild inflammatory reactions. A varying degree of tissue proliferation was seen within the lumen of the shunts with a pattern of cells and connective tissue closely resembling intimal proliferation of stented vessel segments. The extent of pseudointima formation was demonstrated to be time dependent.

### P-53

#### **Development and Characterization of Decellularized Myocardial Tissue Slices**

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**Background:** In the last years Tissue Engineering (TE) focused increasingly on the development of cardiovascular scaffolds. Besides artificial scaffolds there is an increasing demand for native matrices, which are generated by decellularization (DZ). DZ is intended to include the removal of cellular membranes, nucleic acids, lipids, cytoplasmic components and retaining an extracellular matrix (ECM) having as major components collagens and elastins. The aim of this study was to decellularize murine myocardial tissue and thereby to preserve the extracellular matrix.

**Methods:** Murine ventricles were embedded in low-melting agarose and sectioned into 300- $\mu$ m thick slices along the short axis with a microtome. Afterwards the tissue was treated over 3 days with hypotonic Tris-buffer and SDS as well with DNA and RNA nucleases.

**Results:** RNA and DNA could not be detected by PCR-screening. HE staining showed a honeycomb structure. Nuclei could not be detected. Both western blot as well as immunohistochemistry were negative for alpha actinin. Positive staining for fibronectin, collagen IV, nidogen I and laminin I indicated a widely preserved ECM.

**Conclusion:** We could show that the decellularization of myocardial tissue with a vast conservation of the ECM is possible. The decellularized tissue slices could serve as scaffold for the recellularization with adequate cells and are therefore another step in the development of TE techniques in the cardiovascular field.

### P-54

#### **Carvajal/Naxos syndrome secondary to Desmoplakin-dominant mutation is associated with hypo/oligodontia**

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**Introduction:** There is evidence that Carvajal syndrome (wooly hair, palmoplantar keratoderma and dilated cardiomyopathy) and Naxos syndrome (same hair and skin anomalies with fibrofatty cardiomyocytes replacement in the right ventricle) are variable expressions of the same syndrome secondary to mutations in genes encoding proteins of the desmosome. We report here additional signs that might be helpful to cardiac pediatricians to establish the diagnosis.

**Report:** This is a familial case. The proband had 3 episodes of chest pain with transient ST elevation in leads V3-V4 and mild rise of troponin blood concentration. As he was 15 years old, he had left ventricular enlargement (LV diameter 64 mm) with normokinetics (EF 59%) and no coronary anomalies. He had good adaptation to physical activity but numerous premature beats disappearing during physical stress. He had palmoplantar keratoderma, woolly hair and was missing several molars.

A younger brother had 2 fainting episodes as he was 17. He had incomplete RBBB and dilated left ventricle (60 mm diameter at end-diastole) with normokinetics. Four years later, he had shortness of breath with LV at 70 mm and EF at 20%. He had numerous ventricular premature beats and runs of ventricular tachycardia. He received a heart graft. His heart had enlarged ventricles with fibrofatty replacement in anterior and posterior walls of the right ventricle. He had palmoplantar keratoderma, woolly hair and marked oligodontia with only 4 permanent molars and several persisting primary teeth.

The father who experienced several fainting spells had also a dilated cardiomyopathy with the same skin, hair and teeth anomalies.

The desmoplakin (DSP) and plakoglobin (JUP) genes were screened for mutation and a single heterozygous mutation was found in the DSP gene: c.1790C > T, p.Ser597Leu. This residue is conserved across vertebrates. This variant was absent from 100 controls. The mutation was found in the 2 brothers, their father but absent from other family members including the 2 paternal grand-parents.

**Conclusion:** Heterozygous missense mutation in the DSP gene may result in chest pain, fainting episodes, dilated cardiomyopathy in teenagers. The association of woolly hair, palmoplantar keratoderma and/or oligodontia may help in establishing the diagnosis of Carvajal/Naxos disease.

### P-55

#### **Subcutaneous treprostinil in pediatric patients with severe pulmonary hypertension**

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**Background:** Intravenous epoprostenol is first-line treatment for functional class (FC) III/IV Pulmonary Hypertension (PH), but infusion system complications can increase morbi-mortality. Subcutaneous (s.c) Treprostinil (stable prostacyclin analogue) infusion can avoid central catheters complications. Nevertheless, there is limited information about efficacy/safety of this treatment in children.

**Methods:** We report treprostinil use in seven pediatric patients, with mean age of 3.7 + 4.5 years (3 months - 11 years) and weight of 13.6 + 13.9 (3 to 39 kg), with severe pulmonary hypertension (Pulmonary/systemic pressure ratio 104% + 7, PVRI 12,5 + 5,7 WU/m<sup>2</sup>, PVRI/SVRI 1 + 0,1) in functional class III-IV (3,6 + 0,4), in spite of combined treatment with sildenafil and bosentan. The etiologies of PH were: idiopathic

(n = 2), residual PH after CHD surgery (n = 2), Eisenmenger (n = 1), PH associated to metabolic disease (n = 2).

**Results:** Treatment was initiated in seven patients and stopped after two weeks in three (one case due to local intolerance and in two, after definitive diagnosis of metabolic disease with neurologic impairment). In the four patients in which treatment was continued, mean treprostinil dose was  $31 \pm 3.4$  ng/kg/min. Mean follow-up was  $15 \pm 4$  months (11–19 months). At the end of follow-up, FC had improved from III-IV ( $3.6 \pm 0.4$ ) to I-II ( $1.2 \pm 0.5$ ), and in two patients, (3 and 8 months old, with idiopathic and postoperative PH respectively) pulmonary pressure estimated by eco had dropped to near normal levels. Observed side effects were: four days of significant local pain treated with ibuprofen or paracetamol (n = 1), and mild epistaxis (n = 1).

**Conclusions:** 1) Tolerance to s.c. treprostinil was good in most of our patients (Only 1/7 patients (14%) didn't tolerate s.c. treprostinil due to local pain. 2) Subcutaneous treprostinil was safe and effective in PH pediatric patients in FC III/IV in combined treatment with sildenafil and bosentan.

#### P-56

##### **Dilated cardiomyopathy – course and outcome in children**

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**Introduction:** Dilated cardiomyopathy (DCM) is a congenital myocardial disorder resulting end-stage heart failure usually with rapid progression. The aim of study was to provide a detailed description of the etiology, course, and outcome of the DCM in children.

**Methods:** Retrospective analysis of 61 children enrolled in the Hungarian Pediatric DCM Registry between 2005–2010 was performed. Inclusion criteria were defined as decreased left ventricular function (FS < 30%) and/or increased left ventricular end-diastolic diameter (LVEDD > 2SD), which were not caused by any other structural abnormality, or arrhythmia. End points were defined as death, heart transplantation (htx) or implantation of ventricular assist device (VAD). Patient's demographic data, time elapsed from diagnosis to the end points, echocardiographic parameters measured at the time of diagnosis, and left ventricle trabeculation (non-compaction) were investigated.

**Results:** 55 patients were enrolled. Sex 6/5 (m/f); median age at diagnosis was 5.38 years. Non-compaction cardiomyopathy was diagnosed in 15 cases. Familiar inheritance could be proven in 16 cases (29%) involving 13 families. Htx was performed in 11 cases, VAD implantation in 4 cases. The mean time elapsing from diagnosis to htx or VAD application was 3.7 years. In lack of htx 13 patients died. Their average follow-up time from diagnosis to death was 3.39 years. Known etiology was proven only in 1 case (Alström syndrome).

**Conclusions:** 1. Etiology of DCM was not found in most cases. 2. Pediatric DCM has a rapid progression and high need for htx. 3. Familiar cases were found in high percentage of the cases. 4. Due to the high number of idiopathic cases, molecular diagnostic tests are recommended to support early recognition in uncertain cases.

#### P-57

##### **Severe pulmonary hypertension mimicking pulmonary veno-occlusive disease as presenting feature of Non Ketotic Hyperglycinaemia**

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**Introduction:** Nonketotic hyperglycinaemia (NKH) is an autosomal recessive disorder characterized by defective glycine degradation by the mitochondrial glycine cleavage system (GCS), with early or “late-onset” (atypical NKH) neurologic symptoms. The association of Pulmonary Hypertension (PH) to this disease is uncommon, and the characterization of this PH and its response to vasodilator treatment has not yet been reported. We report five infants in which severe PH was the main clinical manifestation of NKH.

**Methods:** Retrospective study of clinical records, echocardiograms, cardiac catheterization, enzymatic studies (liver biopsy), and necropsy findings from five infants diagnosed with PH and NKH.

**Results:** There was history of parental consanguinity in one of the families (three affected siblings) and of a previous son deceased with diagnosis of PH in another. Median age at diagnosis was 3 months (2–4 months). Presenting symptoms: respiratory distress (n = 4), cyanosis (n = 4), poor feeding/vomiting (n = 4), associated intercurrent infection (n = 4). Subsequently, all developed severe respiratory and right heart failure requiring mechanical ventilation and iv inotropes. Only one showed seizures/neurologic impairment. Chest X-ray at admission: normal-sized heart (n = 5), sparse perihilar alveolo-interstitial infiltrates (n = 4). Echocardiography: suprasystemic pulmonary pressure and right-left shunting through foramen ovale (n = 5). Chest angio-CT (n = 3): diffuse ground glass pattern suggestive of pulmonary veno-occlusive disease. Cardiac catheterization (n = 2): normal PCW, median pulmonary/systemic pressure ratio 95%, and pulmonary/systemic resistance ratio 0.8). Median blood and CSF levels of glycine were 1174.4 mcmol/L (649.3–1241.4) and 18.8 mcmol/L (10.9–101.2), respectively, and median GCS levels in liver tissue was  $4.3 \mu\text{kat/Kg prot}$  (0–7.7). Follow-up (median 27 days; range 8–48): after vasodilatory treatments (nitric oxide, sildenafil, bosentan, treprostinil, iloprost, epoprostenol), either alone or in combination, all developed pulmonary edema/haemorrhage, dying of hypoxemic respiratory failure and cardiogenic shock. Necropsy confirmed the diagnosis and CNS involvement, but didn't find anomalies in lung capillary or venous vessels, and only mild/moderate hypertrophy of the muscular layer of intraacinar arterioles.

**Conclusions:** 1) NKH should be ruled out in any infant with PH, even without neurologic symptoms. 2) Vasodilator treatment of PH in NKH induce untreatable pulmonary edema 3) Early diagnosis of NKH-PH could avoid aggressive treatment and allow prenatal diagnosis in subsequent pregnancies.

#### P-58

##### **Results of using of stem cells for treatment of dilated cardiomyopathy at childhood for first six patients**

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**Objective:** Dilated cardiomyopathy is serious disease in pediatric age. Bone marrow derived progenitor cell transplantation becoming a promising method of treatment in adult population and there are describe a few cases in pediatrics. Based on this, we have done the BMCs transplantation in six patients.

**Methods:** The six patients had been admitted for the BMCs transplantation in age at 4 month to 17 year. The diagnosis was

established by Echo, X-ray, laboratory data and endomyocardial biopsy. In May 2009 A. Laci was first in the world to use a percutaneous intramyocardial implantation of BMCs for a 4-month-old child suffering from idiopathic dilated cardiomyopathy, following detailed, multiple observations of the first patient during one year, the decision to use the method in other patients was made. Seventeen to 90 million BMCs were isolated and as a suspension of physiologic saline given to patients by intramyocardial puncture in the interventricular septum. Ejection fraction, NT-proBNP were measured in each patient every two months. The data analysis was made by descriptive and mathematical statistical methods. The statistical significance was determined by t-Test ( $p = 0.05$ ).

**Results:** Six months following transplantation we observed an increase in ejection fraction. The average basal EF was 33.66%. We observed an increase up to 50.25% ( $=7.63$ ,  $p = 0.017415$ ) in a 6-month period. The median basal CTR was 0.63 and it decreased to 0.55 in 6 months. The results were within the confidence interval in all measurements. There were no complications observed during the procedure. The clinical status of patients improved from IV (NYHA) to I-II (NYHA).

**Conclusions:** We see that the intramyocardial administration of bone marrow cells proved to be technically feasible and safe and improves the patients' clinical situation and physical measurements. After short-term evaluation we conclude that stem cell transplantation can be used for the treatment of dilated cardiomyopathy.

#### P-59

##### **15 Year Experience in Pulmonary Hypertension Due to Congenital Heart Disease Before and After Targeted Therapies: The Durability of the Right Ventricle in this disease**

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**Introduction:** Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) is characterized by a severe increase in pulmonary pressure and resistance with relatively preserved RV function and a significantly better prognosis compared to idiopathic PAH. The aim of this study was to examine the 15-year single-center experience in this population before and after use of PAH-targeted therapies.

**Methods:** Between 1995 and 2011, 75 patients, aged at diagnosis  $21 \pm 16$  years, with PAH due to CHD were followed at our center. Diagnoses were: atrial septal defect 3 patients, ventricular/atrioventricular septal defect and/or arterial duct 30, complex CHD 37, Fontan 5. Five patients had Down syndrome while 20 had previous heart surgery. Patients were followed with clinical exam, EKG, echocardiogram, 6-min walk test and cardiorespiratory exercise test every 6 months with laboratory tests every 3 months.

**Results:** Over 15 years (mean follow-up  $8 \pm 5$  years) 96% of patients received PAH-targeted therapy after 2002, initially monotherapy with second therapy added in 8 patients due to clinical worsening (3 patients improved, 5 progressed to death). Most (75%) patients remained stable clinically (WHO Class II) and in exercise capacity, while 14 (19%) deteriorated gradually to Class III. All stable patients had no clinical or echocardiographic signs of RV failure nor did they require treatment for it despite the long follow-up. There were 14 (18.6%) deaths (5 sudden, 9 due to worsening RV failure) at age  $27 \pm 19$  years,  $2.9 \pm 2.3$  years after initiation of therapy. PAH therapy was discontinued in 2 patients after surgical and interventional treatment respectively. All Fontan patients improved significantly with PAH therapy and are expected to discontinue it in the future.

**Conclusions:** Patients with PAH due to CHD, especially in the era of the new PAH-targeted therapies, remain more stable with a better prognosis than idiopathic PAH without significant RV failure over long follow-up periods, possibly because of RV adaptation since fetal life. Still, PAH due to CHD remains a complex disease with significant mortality and morbidity. Close follow-up and individualization of therapy seems critical for these patients' quality of life and long-term survival.

#### P-60

##### **60 patients with Eisenmenger physiology improve clinical status, hemodynamics and pulmonary endothelial function after 24 weeks of Bosentan therapy**

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**Introduction:** Pulmonary vasodilative therapy in pulmonary hypertension of the Eisenmenger physiology has been shown to be safe. We show, in the largest cohort to date, improvement of clinical, hemodynamic and pulmonary endothelial function after 24 weeks of Bosentan therapy.

**Methods:** 60 adult patients (16–51 years, 25m/35f) were recruited and treated for 24 weeks with Bosentan, an endothelin receptor ETA/ETB antagonist. Cardiac catheterisation and acute pulmonary vasoreactivity test, with magnetic resonance and echocardiographic imaging and laboratory markers of cell function (VEGF, BNP, endothelins) were performed before and after Bosentan therapy, which was monitored with 6-MWT, ECHO and clinical status.

**Results:** Clinical status improved (average NYHA  $2.66 \pm 0.59$  to  $2.32 \pm 0.51$ ,  $p < 0.005$ ) as well as 6-MWD ( $65.1 \pm 14.3$  m,  $p < 0.001$ ). Hemodynamics showed a lowered pulmonary vascular resistance at rest, while maximum pulmonary vasodilation with NO/O<sub>2</sub> remained statistically unchanged. Treatment with Bosentan resulted in a significant reduction of pro-BNP ( $p = 0.0011$ ) whereas mid-region-ANP, selectins and cell adhesion molecules remained unchanged. Levels of big endothelin and endothelin increased, and plasma nitrite, nitrate and ADMA showed enhanced NO production.

**Conclusion:** 24 weeks of Bosentan-Therapy in patients with Eisenmenger-Physiology was safe and improved clinical and exercise parameters, and hemodynamics. Specifically, data suggest improved pulmonary endothelial function with endothelin handling and vasodilating capacity of the pulmonary vascular system, demonstrating the beneficial effects of bosentan for the first time in these patients both on the clinical and on the pulmonary vascular cellular level.

#### P-61

##### **Impact of intrauterine and postnatal nutritional determinants on blood pressure in early childhood**

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**Objectives:** To assess maternal nutritional determinants during pregnancy and child postnatal dietary intake, growth and adiposity on blood pressure programming at the age of four years.



**Methods:** A cohort of 109 mother-child pairs was followed from early pregnancy until the children were four years old. Maternal clinical characteristics and dietary intake, recorded in three-day food diaries, were assessed at each trimester of pregnancy. Children's anthropometrics were repeatedly measured, and their dietary intake and blood pressure, using an automated oscillometric DINAMAP ProCare 100, were evaluated at the age of four years.

**Results:** Child systolic blood pressure (BP) was positively associated with maternal carbohydrate intake during pregnancy ( $p = 0.029$ ). The systolic BP was also higher in children exposed to the highest and lowest tertiles of maternal dietary fat intake ( $p = 0.004$ ) and weight gain ( $p = 0.026$ ) during pregnancy, and systolic BP at the 1st trimester ( $p = 0.023$ ), compared to other children. Postnatally, children's dietary fat ( $p = 0.033$ ) and protein intakes ( $p = 0.026$ ), body surface area ( $p = 0.002$ ) and supra-iliac skinfold ( $p = 0.063$ ) were the main factors explaining their systolic BP. The most prominent factors explaining child's diastolic BP were their dietary fiber intake and weight at the age of four. The diastolic BP was lower in children whose fiber intake was in the highest and lowest tertiles, compared to the middle tertile, while weight correlated linearly with diastolic BP ( $r = 0.289$ ,  $p = 0.026$ ).

**Conclusions:** Maternal and child nutritional determinants at the critical period of vascular development may impact on child blood pressure. Interventions are needed focusing especially on balanced dietary intake in mother and child.

#### P-62

##### **Complex Cardiac Prenatal Diagnosis: Continuing or Termination of Pregnancy? A Retrospective Analysis**

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**Objective:** To investigate the complexity of congenital cardiac and extracardiac malformations in fetuses, in whom continuation or termination of pregnancy were considered.

**Methods:** During a five year period (2004–2008) 209 fetuses were diagnosed before the 24th week of gestation having a congenital heart disease (CHD). All parents were counselled twice in a standardized manner by a perinatologist and a foetal cardiologist. The severity level of cardiac and extracardiac malformations were retrospectively categorized using a complexity classification (table 1A/B) by one paediatric cardiologist who was blinded to whether a termination of pregnancy was carried out or not.

Level	Cardiac Malformation
I	No complex CHD, cure possible
II	Complex CHD, cure possible
III	Very complex CHD, only palliation
IV	No therapy feasible

Level	Extracardiac Anomaly
A	None
B	Mild
C	Complex
D	Lethal

**Results:** In 109 (52%) of the 209 fetuses parents opted for termination of pregnancy, at a mean of 21.6 weeks of gestation. Of the 109 terminated pregnancies 101 fetuses (93%) had a cardiac level III/IV and/or an extracardiac level C/D malformation. In other words a level III/IV CHD was present in 49% and a level C/D extracardiac malformation in 59% of the fetuses. 87% of the terminations of fetuses with a CHD level I was associated with a level C/D extracardiac anomaly.

Of the 101 continuing pregnancies 42 fetuses (42%) had a cardiac level III/IV and/or an extracardiac level C/D malformation. In other words 20% had a cardiac level III/IV CHD and 27% an extracardiac level C/D malformation. The complexity level of the CHD or extracardiac malformation was significant higher within the termination group (Fisher's exact test  $P = 0.0001$ ).

**Conclusion:** This is the first report about termination of pregnancy taking into account the severity levels of the foetal congenital heart disease and associated extracardiac malformations. The severity levels of the cardiac and/or extracardiac malformation are significantly higher in the termination group. After thorough counselling it is rare for parents to decide on termination of pregnancy in the absence of severe cardiac and/or extracardiac malformations.

#### P-63

##### **The Impact of Fetal Diagnosis on Duct-Dependent Congenital Heart Lesions**

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**Introduction:** Previous retrospective studies show mixed results for the benefit of fetal diagnosis in congenital heart disease (CHD). The objective of this study was to examine acuity of illness on presentation of prospectively enrolled neonates born with duct-dependent CHD.

**Methods:** Neonates were prospectively enrolled at a single centre and a nested retrospective cohort study examining neonates between January 1, 2009 and October 31, 2009 was conducted. Inclusion criteria were: Neonates with a fetal or postnatal diagnosis of duct-dependent CHD; >36 weeks of gestational age and >2 kg at birth; and CHD requiring intervention within the neonatal period. Exclusion criteria were: major extracardiac congenital or genetic anomalies. The primary outcome was initial lactate at CHD diagnosis in hospital. Other outcomes evaluated were initial pH and creatinine, age of prostaglandin initiation, age of transfer to a tertiary care centre for surgical management, highest dose of prostaglandin, days to initial intervention, and length of ICU stay pre-intervention. Mean values of outcomes were compared using an unpaired t-test. Data are presented as mean with SD.

**Results:** 100 neonates were reviewed; 30 with fetal diagnosis and 28 with postnatal diagnosis fulfilled criteria. Occurrence of transposition of the great arteries and obstructive left and right heart lesions was similar in the 2 groups. The 2 cohorts were also similar in birth weight and gestational age and in APGAR scores. At admission, lactate was significantly lower in the fetal cohort ( $3.6 \pm 2$  vs.  $5.7 \pm 4.4$  mmol/L,  $p < 0.05$ ), while initial pH, creatinine and age of prostaglandin initiation were not statistically different. The fetal cohort received less prostaglandin to maintain ductal patency ( $0.03 \pm 0.04$  vs.  $0.06 \pm 0.04$  mcg/kg/min,  $p = 0.004$ ). Patient age of transfer for surgical management was earlier in the fetal cohort ( $1.1 \pm 0.6$  vs.  $4.9 \pm 6$ ,  $p < 0.001$ ) as was the time of surgical intervention ( $5.5 \pm 3.1$  vs.  $8.8 \pm 7.1$  days,  $p = 0.02$ ) but this did not affect length of ICU stay.

**Conclusions:** Fetal diagnosis of duct-dependent CHD was associated with lower lactates at admission, lower prostaglandin dosages to maintain ductal patency, as well as earlier age of transfer for intervention and earlier intervention. However, this did not shorten the length of ICU stay.

#### P-64

### Diastolic dysfunction in fetuses of Diabetic Mothers, Prenatal two dimensional and Doppler echocardiography study

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**Background:** Pre-gestational diabetes occurs in 0.5%, while gestational diabetes occurs in 1–4%. Fetal hypertrophic cardiomyopathy due to maternal diabetes occurs in up to 35% of fetuses and may cause ventricular diastolic dysfunction, that can be evaluated by echocardiography. Our purpose was to evaluate cardiac diastolic function in fetuses of diabetic mothers using pulmonary veins Doppler flow velocity pattern.

**Methods:** Using trans-thoracic echocardiography in 65 pregnant women, Doppler flow velocity pattern in the pulmonary veins, mitral and tricuspid valves, and M-mode echocardiography measurement of Right and left ventricular free wall, (RV/LV FW), and interventricular septal (IVS) thickness were recorded. All women were divided into three groups: Group 1: included 21 women with pre – gestational diabetes. Group 2: included 25 women with Gestational diabetes. Group 3: included 19 without diabetes. Gestational age was 32–35 wk. All the fetuses were without cardiac or extracardiac malformations.

**Results:** Mean gestational age of the study fetuses was 33 +/- 2 weeks. Pulmonary veins Doppler, interventricular septum and left and right ventricular free wall diameters were recorded in the three groups. Results were in all groups (Group 1 – Pre- Gestational Diabetes, group 2 Gestational Diabetes and group 3 Controls) were respectively as follows: Mean PV S/D ratio was (0.74, 0.64, 0.62): Left ventricular free wall (3.59 mm, 3.52 mm, 2.53 mm) and right ventricular free wall 3.48 mm, 3.57 mm, 2.77 mm.

**Conclusion:** Myocardial hypertrophy in Pregestational and gestational diabetes may cause ventricular dysfunction that can be evaluated by Doppler echocardiography of pulmonary veins. Pulmonary veins Pulsatility are higher in fetuses with normal glycemia.

#### P-65

### Maternal effects of transplacental flecainide for treatment of fetal tachyarrhythmia

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**Background:** Transplacental flecainide is an established treatment of fetal tachyarrhythmias. Its use has been cautioned due to the potential for maternal pro-arrhythmic effects. Previous work suggests that prolongation of the QRS duration (QRSd) >50%, PR interval by >30% and QTc by >15% is an indication of toxic flecainide levels. In pregnant mothers relatively high doses are administered (100 mg t.i.d) necessitating monitoring.

**Methods:** Retrospective review of fetal tachyarrhythmias treated with flecainide from January 1997 to December 2008. Side effects, flecainide levels and ECG data were collected. Automated PR interval and QRSd were manually confirmed. Rate corrected QT interval (QTc) was calculated.

**Results:** 31 cases were identified with persistent SVT (20), intermittent SVT (7) and atrial flutter (4). Median gestation at initiation 32weeks (range: 21–38).

Median trough flecainide level was 410 mcg/l (250–840) within 5days of initiation of treatment (laboratory therapeutic range 200–700 mcg/l). An ECG was performed at the same clinic visit as blood levels. ECG measurements and percentage change from baseline are expressed as median(range) in table 1. In 9 cases PR interval increased >200ms and in 10 cases QTc increased >450 ms reflecting prolongation of QRSd.

Table 1: ECG measurements and percentage change from baseline with flecainide

	PR interval (msec)	% change in PR interval	QRS duration (msec)
Pre-treatment	136 (100–164)		80 (74–104)
Early (2–7 days) with therapeutic flecainide level	160 (122–200)	17 (–3–50)	92 (80–116)
	P < 0.0001		P = 0.0003
At peak flecainide levels	163 (116–218)	24% (0–50)	96 (71–112)
	P < 0.0001		P = 0.0003
	% change in QRS duration	Mean QTc (msec)	% change in mean QTc
Pre-treatment		415 (374–459)	
Early (2–7 days) with therapeutic flecainide level	10% (–2 to 30%)	429 (385–505)	4% (–3 to 19%)
	P = 0.03		
At peak flecainide levels	13% (–4 to 30%)	429 (338–505)	4% (–18 to 27%)
		P = 0.03	

A 30% increase in PR interval from baseline gave a positive predictive value (PPV) for supra-therapeutic flecainide level of 45% and a negative predictive value (NPV) of 91%. QTc increase >15%, PPV = 44%, NPV = 85%. QRSd did not increase >50%.

There was no significant correlation between flecainide levels and PR or QRSd ( $r^2 = 0.118, 0.105$ , respectively).

7(23%) mothers had side effects including: nausea and vomiting (n = 2), visual disturbance (n = 1), palpitations (n = 1), malaise (n = 2), dizziness (n = 3), shortness of breath (n = 1), anxiety (n = 1) and headaches (n = 1). Flecainide levels were supratherapeutic in 2/7 symptomatic patients necessitating dose reduction. Prolongation of the QTc coupled with side effects led to cessation of therapy in 1 patient and reduced dose in 3. There was no maternal mortality or long-term morbidity.

**Conclusion:** There is significant prolongation of the PR interval, QRSd and QTc with flecainide therapy. However, there does not appear to be any correlation between maternal flecainide levels and ECG intervals. The predictive value of ECG changes to predict suprathreshold levels of flecainide appears suboptimal.

#### P-66

### Performance of antenatal diagnosis to detect postnatal coarctation of the aorta

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The aim of this study is to assess the postnatal outcomes of fetuses diagnosed with potential aortic coarctation.

**Material and Methods:** the records of all neonates with antenatal suspicion of coarctation of the aorta were reviewed retrospectively for clinical, echocardiographic data and treatment.

**Results:** Among 78 fetuses diagnosed with abnormal asymmetric ventricles, 35 (44.9%) developed coarctation of the aorta, 42 did not (52.5%) and 2 had hypoplastic left heart (2.6%).

All patients were hospitalized from birth until closure of the ductus arteriosus (2 to days) or Crafoord surgery. The antenatal RV to LV ratio was 1.74 in patients with coarctation of the aorta compared to 1.39 in those without ( $p = 0.018$ ), and the pulmonary artery to ascending aorta ratio was respectively 1.69 versus 1.30 ( $p = 0.0001$ ). The frequency of left superior vena cava was not different between patients with and without postnatal coarctation (29.7 versus 25%). The 2 cases with hypoplastic left heart died postnatally. All the other patient were asymptomatic, and had no symptom of heart failure.

Mean postnatal LV diameter was 15.7 mm, ascending aorta 7.2 mm. None of the patients required prostaglandin infusion or preoperative mechanical ventilation. Crafoord operation was performed at median age of 11.5 days. Median hospital stay was 21.4 days for operated patients.

**Conclusion:** The performance of antenatal diagnosis to detect postnatal coarctation of the aorta remains low. However RV to LV and PA to AO ratios may help to ameliorate specificity. Despite high rate of false positive, antenatal diagnosis allows to avoid postnatal acute heart failure and improve early prognosis.

### P-67

#### **Prediction of biventricular versus univentricular circulation in fetal diagnosis**

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**Introduction:** An important aspect of fetal diagnosis is prediction of univentricular (UV) versus biventricular (BV) circulation, especially if diagnosis is made <22 weeks when termination of pregnancy (TOP) is possible. We sought to determine the accuracy of this prediction in our experience.

**Methods:** 85 consecutive fetuses with prenatal diagnosis of complex heart malformations (need for intervention <1 year) were eligible but only those 68 who had a postmortem (PM) or postnatal diagnosis were included. The prediction by the attending fetal cardiologist at the time of the first fetal echocardiogram (UV or BV) was compared to the fetal PM or the hemodynamics at the last postnatal follow-up. A prediction of possible UV was analyzed as UV. If there was more than one examination before 22 weeks the final judgement was used.

**Results:** In no case was the prediction different at fetal follow-up compared to first impression.

**Top:** The fetal prediction was UV in 20 and BV in 4. This was confirmed in all.

**Fetal deaths:** In 1 of 5 the fetal prediction was UV which was confirmed. Four were predicted to be BV, confirmed in 2, but 2 could also have been UV according to PM (1 AVSD, 1 PA/IVS).

**Liveborns:** In 15 of 39 UV was predicted. Twelve had UV and 3 had BV (1 AVSD and 2 with Coarctation of the Aorta). Twenty-four were predicted to be BV but 4 had UV (2 AVSD, 1 DORV/PA, 1 DILV/TGA).

**Diagnosis:** <22 weeks.

Twenty-three of 33 were predicted to be UV and 10 BV. Prediction was incorrect in 2. Both had AVSD predicted to be BV but after live birth one had UV. The other fetus died and PM showed probable UV.

**Conclusion:** The prediction of UV versus BV was correct in 59 of 68 cases and in 31 of 33 diagnosed <22 weeks. Four of 9 with incorrect prediction had AVSD and 2 had Coarctation of the Aorta. In no case of TOP was the prediction incorrect as judged from PM. However PM may not have correctly reflected the postnatal situation in every case had the pregnancy continued.

### P-68

#### **Risk for isolated ventricular septal defect in euploid fetuses with borderline or increased first trimester nuchal translucency**

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**Basics:** The association of an increased nuchal translucency (NT) in the first trimester with a higher prevalence of fetal complex structural cardiac defects, in the absence of chromosomal abnormalities, has long being reported. The purpose of this study was to test the hypothesis that there is association between a borderline or increased NT (2 mm or more) in 11 + 0–13 + 6 weeks euploid fetuses and a later diagnosis of isolated VSD.

**Methods:** A case-control study was designed. 5464 second or third-trimester consecutive fetuses without extracardiac abnormalities and no cardiac anomalies other than a VSD were assessed in a period of two years. NT had been obtained between 11 + 0 and 13 + 6 weeks of gestation. A fetal echocardiogram and a morphological scan were performed, searching for the diagnosis of VSD and to exclude associated cardiac and noncardiac malformations. Fetuses with an altered karyotype or a postnatal diagnosis of chromosome abnormalities were excluded. Statistical analysis used Fisher's exact test and ROC curves.

**Results:** Mean maternal age was  $32 \pm 5$  years (21–42 years) and gestational age at the time of the fetal echocardiogram was  $25 \pm 6$  weeks (19–31 weeks). Mean NT was 2.2 in fetuses with VSD and 1.4 without. Out of 319 fetuses with a NT of 2 mm or more, 67 had a VSD (52 muscular and 15 perimembranous) (21%), while in only 115 out of 5180 fetuses with a NT < 2.0 mm a VSD was detected (86 muscular and 29 perimembranous) (2.2%) ( $p < 0.0005$ ), being the relative risk = 9.3 (99% CI: 6.5–13.5). A ROC curve determined the cut-off value of NT (2.0), with a sensitivity of 48.3% and a specificity of 91.4%, with an area under the curve = 0.695 ( $p < 0.0001$ ).

**Conclusions:** Fetuses without chromosome abnormalities with a first trimester NT of 2 mm or more have an 8.3-fold increase in the risk of presenting an isolated ventricular septal defect. We speculate that the defects could have been larger and functionally important in the first trimester, increasing the NT as a consequence of hemodynamic overload and gradually decreasing its diameter until the second and third trimesters. This knowledge may have implications in prenatal management and counseling.

### P-69

#### **The fetal heart in monochorionic twin pregnancies: how much are we missing?**

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**Introduction:** Twin pregnancies have an increased risk of cardiac structural and functional abnormalities compared with singleton pregnancies. This risk appears to be even greater when we refer to monochorionic pregnancies. The aim of this retrospective study is to review the incidence of cardiac disease in a population of monochorionic twins.



**Methods:** Retrospective review of all monochorionic twin pregnancies referred for a fetal cardiac scan in our institution. Data about gestational age and cardiac diagnosis was reviewed. Chorionicity and type of cordal insertion was confirmed through the histologic analysis of the placenta.

**Results:** Between January 2007 and December 2010, 3803 fetal cardiac scans were performed in our department. Of these, 166 were monochorionic twin pregnancies. In 12 (7.2%) of these there was a diagnosis of congenital heart disease in one of the twins. The cardiac diagnosis was: simple complete transposition of the great arteries (2), transposition of the great arteries with a ventricular septal defect (2), aortic stenosis and coarctation of the aorta (1), right isomerism with aortic atresia (1), ventricular septal defect (3), pulmonary valve stenosis (1), pulmonary atresia with intact ventricular septum (2). In the 3 cases of pulmonary stenosis and atresia, a diagnosis of twin to twin transfusion syndrome (TTTS) was also made by echocardiography. The cardiac malformations were diagnosed in the receptor twin, and in all 3 cases laser therapy was successfully performed. In 10 cases, the type of cord insertion was assessed. The most frequent type of cord insertion was central (12), followed by marginal (4), and velamentous (3). In one case information about cord insertion was not available.

**Conclusions:** During the period of the study, the incidence of cardiac disease in our population of monochorionic twins appears significant. The reason for these different cardiac malformations is not yet clear, but one may speculate that hemodynamic imbalance based on a single placenta with anastomoses is a frequent event and may contribute to the “remodeling” of cardiac anatomy.

#### P-70

##### **Obstetric management and time to first intervention in cases of fetal congenital heart disease delivered in a tertiary centre**

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**Introduction:** Following prenatal diagnosis of congenital heart disease (CHD), delivery is arranged at our tertiary cardiac centre if early intervention is anticipated. This often necessitates planned induction of labour, which is associated with an increased rate of instrumental delivery or emergency Caesarean section (CS).

**Aim:** To determine the timing of neonatal intervention in babies with antenatally diagnosed CHD delivered in our centre and the impact on obstetric management.

**Methods:** Between 1st Jan 2008 and 31st December 2009, 205 women delivered 206 babies with prenatally diagnosed CHD at our tertiary centre. Data were obtained by retrospective review of fetal, paediatric and midwifery databases.

**Results:** 98 women (48%) lived > 24 km from our center. Of 107 patients living within 24 km, 21 (19.6%) were already booked for delivery at our hospital regardless of fetal CHD. 10/205 (4.9%) delivered at our centre following parental or local hospital request.

119 babies (58%) needed catheter or surgical intervention during their initial postnatal stay: 31% within 48 hours and 42.9% within 72 hours. Median time to first intervention was 4 days (IQR 2–8). Babies with hypoplastic left heart (median 3, IQR 2–6.5), transposition of the great arteries (median 1, IQR 0–5) and arrhythmia (median 0.5, IQR 0–1) had significantly earlier time to first intervention (median 4–11 days;  $p = 0.001$ ). Of other

conditions 11/27 (40.7%) with Tetralogy of Fallot required intervention during their hospital stay, 5 (18.5%) within 72 hours. All 29 with coarctation required intervention; 2 (6.9%) within 72 hours. 12/14 (85.7%) with pulmonary atresia required intervention; 1 (7.1%) within 72 hours. 28/205 babies (14%) died within 30 days.

There was no difference in emergency CS rate following spontaneous labour (39%) compared to following induction of labour (42%) ( $p = 0.74$ ) or between patients living within 24 km of the hospital (51%) compared to those living further away (46%)  $p = 0.75$ .

**Conclusion:** Duct dependent lesions require early intervention and delivery in or near a tertiary centre is recommended. However, this is not necessary in all major forms of CHD. Further studies are needed to accurately identify cases requiring early intervention. There is no significant difference in the obstetric outcome by delivering mothers at the tertiary unit.

#### P-71

##### **Foetal dysfunction of the arterial duct: clinical spectrum and outcome**

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**Background:** premature foetal duct closure cause right heart & pulmonary damage. Outcome of milder dysfunction (partial or transient constriction, aneurysm formation) needs to be defined.

**Methods:** retrospective review; foetal (942) and neonatal echo databases searched for evidence of prenatal ductal dysfunction (1998–2010). Prenatal inclusion criteria were closure – constriction – aneurysm of arterial duct. Postnatal (<14d) inclusion criteria were excessive RV hypertrophy; cyanosis by atrial R < L, and/or abnormal findings duct.

**Results:** 26 patients were identified. 12 prenatally (gestational age 25.7w (21–37w, incidence: 12/942 = 1.3% abnormal scans), 13 postnatally (age D2, range D0–11).

In the foetal cases the duct was closed (9), abnormally small (2), aneurysmal (1). 6 patients presented at birth with significant cyanosis without duct. 5 mothers had taken NSAID during pregnancy. Patients had RVH (25), bipartite RV (16), RV aneurysm (1), significant TR (23), hydropericardium (2) and hydrops (1); PS (6) and PR (22) ranging from mild to “agenesis” of pulmonary valve (3); dilation of pulmonary trunk (6) and branch pulmonary arteries, compression of airways with “fluid-trapping” and microcystic malformation of lungs (1); fetal suprasystemic pulmonary hypertension (5); 2 pt had a thrombosed aneurysm of the duct. In 6 patients premature delivery was chosen to avoid further intra-uterine damage of the right heart & lungs. Neonatal treatment varied from observation (12), ventilation with pulmonary vasodilators (8) and ECMO (1); resection of a thrombosed aneurysmal duct as the thrombus was occluding the left pulmonary artery (2); 3 patients had balloon dilation PS. 3 patients died in the neonatal period because of respiratory insufficiency. Late treatment: balloon dilation PS (2), and homograft reconstruction with PA plasty at 4 years for late compression of coronary artery (1). 2 pts have mild psychomotor delayed development; non-compaction cardiomyopathy (1).

**Conclusions:** Fetal dysfunction of the arterial duct can stress at different fetal ages many different levels of the right heart and pulmonary circulation, resulting in a very wide range of secondary pathology. The clinical outcome ranges from normal to death; neonatal death was due to lung damage. Premature delivery might be indicated in selected patients.

## P-72

**Function of right ventricle in a recipient twin in twin to twin transfusion syndrome**

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**Introduction:** Twin to twin transfusion syndrome (TTTS) is a condition affecting around 10% of monochorionic diamniotic twin pregnancies. Various echocardiographic changes have been described in the recipient twin. The disease affects function of recipient's twin right ventricle due to volume overload, increased afterload and myocardium remodeling.

**Methods:** We reviewed our computer database on fetal echocardiographic exams of twins between 2002 and 2010. 43 pairs of twins with TTTS were identified. They were divided into groups according to Quintero staging system. For each recipient twin cardiac size, myocardial thickness, contractility, flow through tricuspid and pulmonary valves and right ventricle myocardium performance index (MPI) were evaluated.

**Results:** There were 19% (8pts) in Quintero stage 1 (Q1), 21% (9pts) in Q2, 42% (18pts) in Q3, 16% (7pts) in Q4, 2% (1pts) in Q5. Fetal heart was enlarged in 56% (24pts). Myocardial thickness was increased in 58% (25pts). Shortening fraction was decreased in 60% (26pts). Tricuspid regurgitation was present in 81% (35pts) and in 16% (7pts) fusion on right ventricle inflow was observed. Pulmonary stenosis was described in 14% (6pts). Mean MPI for this group was  $0,662 \pm 0,25$ , while in the group of healthy monochorionic twins  $0,407 \pm 0,08$ . In the table distribution of echocardiographic changes according to Quintero stages is shown.

	Q1	Q2	Q3	Q4
Heart enlarged (%)	50%	33%	50%	100%
Myocardium thickened (%)	63%	67%	39%	86%
Shortening fraction decreased (%)	63%	56%	56%	86%
Tricuspid regurgitation (%)	88%	89%	78%	100%
Monophasic RV filling (%)	0%	11%	17%	43%
Functional Pulmonary atresia (%)	13%	0%	6%	57%

**Conclusions:** Function of right ventricle is affected by TTTS in most recipient twins. Serious cardiac function alterations are presents regardless of Quintero stage, so cardiac changes are more important to evaluate twin condition in TTTS. Echocardiographic examination should be performed in all cases of suspected TTTS.

## P-73

**Pulmonary atresia with intact ventricular septum; second heart field derived developmental clues for myocardial and coronary artery pathology**

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**Introduction:** Pulmonary atresia with intact ventricular septum (PAIVS) is a severe cardiac malformation that is ductus dependent and needs immediate postnatal care. The pathology concerns variable degrees of hypoplasia and hypertrophy of the right ventricle including tricuspid valve pathology. The pulmonary valve is atretic which may have been preceded, in the fetal stage,

by pulmonary stenosis. Severe coronary anomalies, characterized by ventriculo-coronary arterial communications (VCAC) are found in 30% of the patients. Current cardiac developmental data show that the right ventricular (RV) myocardium is derived from the second heart field (SHF). Its anterior part contributes this myocardium to the RV outflow tract and is also employed by the neural crest cells (NCC) that enter the heart. The posterior SHF provides the epicardium derived cells (EPDC) that are important for myocardial wall compaction and formation of the coronary vasculature. We hypothesize that PAIVS without VCACs is primarily based on anterior SHF directed outflow tract septation anomalies including NCCs, while PAIVS with VCACs have a major problem related to an EPDC contribution.

**Methods:** We studied human fetal and neonatal postmortem specimen with known clinical diagnostic data. For our immunohistopathological evaluation we used antibodies that reveal a possible role for EPDC as well as myocardial and vascular wall differentiation markers.

**Results:** We showed that the presence of coronary arterial wall pathology in VCACs and concurrent coronary arterial interruptions could be present already in the fetal stage and predisposed in these cases to development from pulmonary stenosis to pulmonary atresia. Normal main coronary arteries in combination with endocardial fibroelastosis of the RV were the hallmark of a second category. We are currently evaluating a difference in myocardial pathology between both groups.

**Conclusions:** The above findings supported our hypothesis that PAIVS with VCAC form a primary disease that is distinct from the cases with PAIVS and endocardial fibroelastosis and normal main coronary arteries. Developmentally they can be considered as two different diseases that might need separate treatment protocols. Study of family history might reveal whether PAIVS with and without VCACs are genetically linked which can direct further developmental studies.

## P-74

**Fetal Cardiomyopathy multicentre study: aetiology and clinical outcome**

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**Objectives:** Although prenatal diagnosis of cardiomyopathy (CM) is rare, it has a high mortality and morbidity. The aim of our study is to describe the underlying causes, hemodynamic and echocardiographic findings and the outcome of these patients.

**Methods:** Multicentre retrospective study between January 2007 and November 2010 at 6 tertiary Spanish centres. We included 45 fetuses with dilated (DCM), hypertrophic (HCM) or Non Compaction (NCCM) cardiomyopathy. We excluded fetuses with congenital heart disease, arrhythmias, twin-twin transfusion and hypervolemic causes. Mean gestational age at diagnosis was 26 weeks (range 17–38 GW), with 2 twin pregnancies. Post-natal follow-up or necropsy was achieved in 92% of cases.

**Results:** DCM was diagnosed in 26 cases including 3 congenital infections, 4 endocardial fibroelastosis due to maternal anti-Ro/La and 19 idiopathic causes. During postnatal life 4 dilated cases turned out to be NCCM. 17 fetuses had HCM: 1 with familial hypertrophy, 4 associated with maternal diabetes, 3 with Noonan Syndrome, 1 Pompe, 1 with polichystic kidneys and 7 with

idiopathic hypertrophy. Intrauterine diagnosis of NCCM was found in 2 cases. Fetal echocardiographic findings were: cardiomegaly 80%, abnormal Ductus Venosus/inferior vena cava flow 63%, tricuspid or mitral regurgitation 56%, systolic dysfunction 43%, diastolic dysfunction 33%, prolonged isovolumetric relaxation time and Tei Index 71% (when measured). Hydrops was found in 38% of cases. Termination of pregnancy and intrauterine death occurred in 20% and 9% of patients. Transplacental medication was administered to 7% of fetuses. Fetuses with tricuspid/mitral regurgitation, prolonged Tei index and anomalous venous flow had a higher mortality. Out of the 23 pregnancies that continued, 16% died perinatally, 8% died before 2 years old, and 4% (2) were included in heart transplant program (1 died on Berlin Heart). Overall survival was 44% (56% in non-hydrops fetuses compared to 21% in hydrops fetuses), 47% of the survivors are under oral medication.

**Conclusions:** Fetal cardiomyopathy has a high prenatal and postnatal mortality. Hydrops, tricuspid/mitral regurgitation and abnormal venous flow pattern are strong predictors of poor outcome. Diagnosis of NCCM is feasible during fetal life.

#### P-75

##### **Foetal dysrhythmias: a study in nine Spanish centres**

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**Objective:** To review current management and outcome of foetal arrhythmias in Spain.

**Methods:** Retrospective multicentre study: analysis of all fetuses with dysrhythmia, as diagnosed by echocardiography, at 9 Spanish Hospitals between January 2008 and September 2010.

**Results:** 123 pregnant women fulfilled the study criteria. The mean gestational age at diagnosis was 29 weeks.

The most common type of foetal arrhythmia was premature atrial contractions, leading to a benign prognosis.

52 of the fetuses studied showed sustained dysrhythmia of which 16 exhibited complete atrioventricular block (CAVB), 4 atrial flutter (AF) and 32 re-entrant tachycardia involving the atrioventricular node (SVT).

**CAVB:** One third of the patients presented with hydrops. 62% of the fetuses were treated with dexamethasone (18% in association with sympathomimetics). Dexamethasone did not improve the degree of blockage in any of the patients. The rate of mortality, mostly associated with ventricular dysfunction, was 21%.

**AF:** There was no case with hydrops. Two fetuses were treated successfully intrauterine (1 with digoxine and 1 with sotalol) and two fetuses diagnosed near term were cardioverted after delivery.

**SVT:** 21 non-hydrops fetuses. Prenatal control of the tachycardia was achieved in 86% of the treated cases: digoxin monotherapy converted 50% of them, second line treatment were sotalol and flecainide. There was no mortality in this group.

**SVT:** 11 hydrops fetuses: management strategies were highly diverse: digoxine, propranolol, flecainide, sotalol, amiodarone and direct foetal administration of adenosine. Six patients died (54%): 1 after delivery; 3 intrauterine very shortly after being evaluated at

the tertiary care centre (mean time: 2 days); 2 intrauterine in spite of being successfully cardioverted to sinus rhythm (1 with sotalol, 1 with flecainide).

**Conclusions:** The management of foetal arrhythmia is still controversial, as shown by the diversity of treatments used in our study. Hydrops fetuses with SVT showed significant mortality in our population, which calls for further studies and unification of criteria. This study is the first step towards the development of a common management strategy in Spain to optimize the outcome of foetal dysrhythmia.

#### P-76

##### **Prenatal diagnosis of pulmonary atresia and intact ventricular septum – can we predict rationale for prenatal intervention?**

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**Objective:** To evaluate anatomy, intrauterine evolution and outcome of fetuses with pulmonary atresia and intact ventricular septum (PA&IVS) diagnosed in tertiary center for fetal cardiology and look for the indication for prenatal intervention.

**Method:** 28 fetuses with confirmed PA&IVS between 1995–2010. Anatomy of the RV, pulmonary artery, tricuspid regurgitation (TR), sinusoids and neonatal outcome was evaluated. Measurements of the right and left heart structures were repeated, Z-scores were calculated. Fetal measurements were compared with the results of treatment.

**Results:** The mean age of diagnosis was 27 +/- 6 weeks, 10 was made before 24 weeks. In 16 karyotype was normal, not signs of chromosomal anomalies in others. One pregnancy was terminated, the fetus had skeletal dysplasia. There was one late intrauterine death. 3 neonates were born prematurely, two were hydropsic, all 3 died in the neonatal intensive care unit. 23 children were treated. Radiofrequency was not performed. In two neonates critical pulmonary atresia was diagnosed and pulmonary valvuloplasty following by BT shut was performed. In the rest 21 BT shunt was done in the neonatal period. 10 children died after surgical treatment: 7 after BT shunt, all had sinusoids, 3-additional TR, 1 died after hemi-Fontan and 2 after Glenn. None of them had TR. 13 children survived, 11 with holosystolic TR with high velocity. Survival rate was 46% for all, 50% for live-born, 56% for operated neonates. RVOT opening was performed in 2, both had Z-score of TV within normal limits. There were not children after biventricular operation. There was not significant difference in Z-scores of the diameter of TV annulus and the size of RV between fetuses with and without TR, and those who died and survived. The medium value of Z-scores for RV diameters about -4, medium Z-score for the pulmonary artery branches was normal.

**Conclusions:** The prognosis for fetuses with AP&IVS in our experience is poor. Prematurity is the important risk factor for neonatal death. Due to the lack of biventricular repair and high rate of death after surgical treatment prenatal intervention should be considered to change the prenatal natural history of this severe disease, even if the right ventricle is hypoplastic.



**P-77****A decade of antenatally diagnosed unbalanced ventricles – comparing fetal outcomes with a postnatal cohort**

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**Objectives:** To compare presentation, management and outcome in a cohort of antenatally (AN) and postnatally (PN) diagnosed patients with unbalanced ventricular chambers referred to a single tertiary fetal cardiac unit and tertiary paediatric cardiac surgical centre.

**Methods:** Retrospective review of the fetal database and case-notes of patients seen between 2001 and 2010 at Liverpool Women's Hospital and Alder Hey Liverpool Children's Hospital. All fetuses diagnosed with unbalanced ventricles, as well as those with borderline ventricular chambers were included. Among postnatally diagnosed babies, only confirmed unbalanced chambers were included.

**Results:** 256 patients were diagnosed with complex cardiac lesions with unbalanced ventricles (AN = 207; PN = 49). The annual antenatal detection rates remained constant during the period at 80% (SD +/- 10%). Reasons for antenatal referral were predominantly abnormal four chamber views on routine screening (96%) with approximately 1% each for positive family history, associated anomalies and abnormal outflow tracts. The gestation at initial antenatal referral was less than 22 weeks in 46% (mean +/- SD 10%) – 44% of these opted for termination. When antenatal referral was made later than 22 weeks, similar proportion of cases were terminated (mean 40%). This trend was consistent over the last decade. On a yearly basis, 42% (mean ± SD 14%) of pregnancies diagnosed with ventricular size imbalance underwent termination or ended in spontaneous fetal demise. Among liveborn babies, 69% in the antenatal group and 90% in the postnatal group underwent active intervention. Antenatal detection did not influence longterm survival in complex unbalanced ventricles.

**Conclusions:** In our region, antenatal detection of complex cardiac lesions with unbalanced ventricular size is high. Termination rates have stayed constant, irrespective of the gestation of antenatal referral. In our study, active management was less likely pursued in the antenatally diagnosed group (AN = 69%; PN = 90%). Survival following active management did not significantly differ between the antenatally and postnatally diagnosed groups.

**P-78****Fetal supraventricular extrasystolies in a referral centre of fetal cardiology**

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**Objective:** To review the cases of fetal supraventricular extrasystolies (SVEx) in a referral centre of fetal cardiology.

**Material and method:** We analyzed the results of echocardiographic examinations of 200 fetuses with supraventricular extrasystolies. Gestational age at diagnosis was 18 to 41 weeks (mean 31 ± 5 weeks).

**Results:** 195 women were in single pregnancy. In 5 cases SVEx were diagnosed in one fetus of twin pregnancy. In 103 (51,5%) cases SVEx were multiple and in 97 (48,5%) single. 188 (94%) fetuses had normal heart anatomy. Heart defects were diagnosed

in 12 (6%) cases (VSD – 5, ISVC – 2, TOF – 1, AT- 1, DORV – 1, AVSD – 1, TGA – 1). 64 (32%) fetuses with SVEx had atrial septal aneurysm. 11 (5,5%) cases of SVEx with normal heart anatomy were complicated by tachyarrhythmia: 10 (5%) fetuses developed intermittent supraventricular tachycardia and one (0,5%) intermittent atrial flutter. In remaining 189 fetuses the SVEx resolved spontaneously prior to delivery in 137 (72%) cases. In 15 (7,5%) fetuses multiple blocked atrial ectopic beats caused low ventricular rate below 100 bpm.

15 (7,5%) women had tocolysis with fenoterol when fetal SVEx were diagnosed.

**Conclusions:** SVEx are usually a benign condition, which resolves spontaneously prior to delivery. However in 5,5% they can progress to tachyarrhythmia and in 5% may be associated with heart defects. Atrial septal aneurysm was observed in 32% fetuses with SVEx.

**P-79****Serum adiponectin, resistin levels, insulin resistance and cardiac changes in obese and overweight children**

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**Aim:** The aim of study is to evaluate the cardiac changes in children with obesity, asses serum adiponectin and resistin levels, and insulin resistance in childhood obesity.

**Material and methods:** Seventy one obese children whom body mass index > 97 percentile (44 boys, 27 girls), 24 overweight children whom body mass index between the 85–97 percentile (6 boys, 18 girls) and 40 age and sex matched control children were selected to study. Systolic, diastolic functions of left ventricle, left ventricular diameters, wall thickness and left ventricular mass, Tei index, aortic stiffness index were measured by using conventional 2D, colour coded echocardiography and tissue doppler echocardiography in obese children, overweight children and control group. After echocardiographic examination blood samples were taken and serum adiponectin and resistin levels were measured with ELISA. Fasting insulin levels measured with Immunoassay. After Shapiro-Wilk normality test results were assessed with chi-square test, unpaired t test, Mann-Whitney U test, ANOVA test and linear regression test by using SPSS for version 13,0. All p value <0.05 were considered statistically significant.

**Results:** In echocardiographic evaluation, obese and overweight children have higher left ventricular mass values compare to control group (p < 0.01). In conventional echocardiography and tissue doppler echocardiography were showed diastolic abnormalities such as relaxation abnormality in obese and overweight children. Hypertension was determined in 18 patients in obese group and 8 patient in overweight group. Serum adiponectin and resistin levels were significantly lower in obese and overweight group p < 0.05). There was statistically significant correlation between the serum resistin levels, fasting insulin and left ventricular mass in obese group. There was not statistically significant correlation between the adiponectin levels and cardiac parameters. Serum resistin levels and systolic blood pressure found to be positive affecting factors to left ventricular mass in linear regression analysis.

**Conclusion:** This study demonstrated that obese and overweight children have increased left ventricular dimensions, left ventricular

mass and showed diastolic changes in obese and overweight children. Obese and overweight children have significantly lower serum adiponectin and resistin levels than control group. Systolic blood pressure and resistin levels are important factors influence the cardiac remodelling in childhood obesity.

#### P-80

##### **Assessment of vascular remodelling after the Fontan procedure using a novel very high resolution ultrasound method: Arterial wall thinning and venous thickening in late follow up**

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**Objectives and Background:** The Fontan circulation is associated with an increased central venous pressure, decreased ventricular preload and increased afterload. We postulated that these central hemodynamic abnormalities would have consequences for the structural and functional properties of the peripheral arteries and veins.

**Methods:** We prospectively examined venous and arterial wall morphology by very-high resolution ultrasound (25–55 MHz), and function by conventional vascular ultrasound (flow-mediated dilatation, FMD) and applanation tonometry (pulse wave velocity; PWV) in 28 patients after the Fontan procedure (age  $14.8 \pm 1.3$  years) and 54 age-matched controls.

**Results:** The lumen dimension was reduced in Fontans patients compared with controls in the common carotid, brachial, radial, and femoral arteries. The common carotid, brachial, radial, ulnar, femoral and dorsal tibial artery intima-media thickness (IMT), and brachial, ulnar, and femoral artery adventitial thickness were decreased, while the cubital and dorsal tibial vein IMT was increased in Fontans. FMD, abdominal aortic stiffness and carotid-femoral PWV were similar, while carotid-radial artery PWV was increased in Fontans.

**Conclusions:** The Fontan circulation is associated with significant arterial and venous remodelling, presumably reflecting abnormalities of central hemodynamics. These novel data may be of clinical importance in the circulatory management as well as in the understanding of the early pathogenesis of vasculopathy in patients after the Fontan procedure.

#### P-81

##### **Assessment of Vascular Phenotype Using a Novel Very High Frequency Ultrasound Technique in Adolescents after Aortic Coarctation Repair and/or Stent Implantation: Relationship to Central Hemodynamics and Left Ventricular Mass**

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**Background:** Coarctation of the aorta (CoA) is associated with abnormal vascular function, increased blood pressure (BP) and premature cardiovascular disease even after successful intervention. We postulated that residual abnormalities of arterial structure and function and left ventricular (LV) mass are directly related to BP.

**Methods:** Thirty-six youths with CoA (age  $16 \pm 1$  years; neonatal surgery only  $n = 16$  and/or stent implantation in childhood or adolescence  $n = 20$ ) and 37 age-matched controls were prospectively examined by very-high resolution vascular ultrasound imaging (25–55 MHz), echocardiography and applanation tonometry.

**Results:** CoA was associated with increased right arm systolic BP ( $p < 0.001$ ), intima-media thickness (IMT) in the common carotid ( $p < 0.001$ ), right brachial ( $p < 0.05$ ) and radial ( $p < 0.05$ ) arteries and ascending aortic stiffness ( $p < 0.05$ ). Carotid IMT correlated positively with age at first intervention ( $r = 0.36$ ,  $p < 0.05$ ). With left subclavian flap type repair, left arm systolic BP ( $p < 0.001$ ) and left brachial ( $p < 0.001$ ), radial ( $p < 0.001$ ) and ulnar ( $p < 0.05$ ) arterial IMTs were all reduced. When adjusted for BP, body mass index, age and gender, only carotid IMT ( $p < 0.001$ ) and LV-mass ( $p = 0.013$ ) of stented patients, as well as left arm arterial IMTs ( $p < 0.01$ ) in subclavian flap type repair patients remained different from controls. No differences between CoA and controls were found for arterial adventitial thicknesses, lumen dimensions, thigh systolic BP, abdominal aorta and carotid stiffness and right carotid to radial pulse wave velocity.

**Conclusion:** CoA is associated with increased preductal arterial IMT, LV mass and ascending aortic stiffness. The more pronounced findings after CoA stent implantation are likely related to the later age of intervention.

#### P-82

##### **Serum Homocysteine, Asymmetrical Dimethylarginine And Nitric Oxide Levels In Children With Pulmonary Arterial Hypertension**

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**Introduction:** This study has been designed to determine homocysteine, asymmetric dimethyl arginine (ADMA) and nitric oxide (NO) levels in patients with congenital heart defects (CHD) having pulmonary arterial hypertension (PAH) and healthy controls and to define the relationship of these markers with echocardiographic and catheter angiographic findings.

**Materials and methods:** A total of 70 cases including 30 patients with PAH, 20 patients with CHD having left to right shunt without PAH and 20 healthy controls were included in the study. Homocysteine, ADMA and NO levels were determined in plasma samples by ELISA.

**Results:** Homocysteine and ADMA levels were higher in PAH group when compared to others whereas there was no difference in between the groups regarding NO levels. The cyanotic patients in PAH group had significantly elevated homocysteine levels when compared to acyanotics. No correlation was shown in between echocardiographic and hemodynamic parameters and homocysteine, ADMA and NO levels of patients with PAH.

**Conclusion:** We concluded that in the patient group with PAH, increased homocysteine and ADMA levels could have been contributed to development of PAH in association with other factors or might have increased as a result of PAH proving that in either case these parameters can be used as biomarkers of PAH. Key words: Pulmonary arterial hypertension, homocysteine, ADMA, NO.

## P-83

**Impact of Right Ventricular Volume Change on Exercise Capacity: A Comparison of Repaired Tetralogy of Fallot and Ebstein's Anomaly**

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**Background:** Patients after surgical repair of tetralogy of Fallot (rTOF) often develop pulmonary regurgitation, which leads to right ventricular enlargement. This is also observed in patients with Ebstein's anomaly due to tricuspid valve regurgitation. In the present study we compared the recovery of peak oxygen uptake (peak VO<sub>2</sub>) and right ventricular enddiastolic volume (RVEDV) after surgery for pulmonary or tricuspid regurgitation. **Patients and Methods:** We studied 10 patients with rTOF and pulmonary regurgitation (mean age [±SD] 20 ± 7 years) without significant pulmonary stenosis and 10 Patients with Ebstein's anomaly (39 ± 19 years). Patients with rTOF underwent surgical pulmonary valve replacement and patients with Ebstein's anomaly underwent surgery for tricuspid regurgitation. Right ventricular size was assessed with cardiovascular magnetic resonance (CMR). Cardiopulmonary exercise performance was evaluated by determination of the peak VO<sub>2</sub>. CMR and exercise testing were performed in both groups prior and 6–30 months after the operation.

**Results:** After surgical valve repair, there was a significant decrease in the right ventricular enddiastolic volume (RVEDV) in both groups (p<sub>Ebstein</sub> = .007; p<sub>rTOF</sub> = .005) [Table 1]. Peak VO<sub>2</sub> did not significantly change in either group (p<sub>Ebstein</sub> = .38; p<sub>rTOF</sub> = .54) [Table 1].

**Conclusion:** After surgical treatment of pulmonary and tricuspid regurgitation, right ventricular size decreased in patients with rTOF and Ebstein's anomaly. Although RV volume load and RV enddiastolic volume were significantly reduced after surgery, a significant impact on peak VO<sub>2</sub> was not detected in both groups.

Table 1: Mean RVED and mean peak VO<sub>2</sub> values of patients with rTOF and Ebstein, pre- and post-surgery, respectively.

	Ebstein's anomaly	rTOF
RVEDV (ml/m <sup>2</sup> ) pre-surgery	190 ± 83	174 ± 30
RVEDV (ml/m <sup>2</sup> ) post-surgery	121 ± 60	120 ± 24
Peak VO <sub>2</sub> (ml/kg/min) pre-surgery	20.6 ± 4.8	29.8 ± 7.0
Peak VO <sub>2</sub> (ml/kg/min) post-surgery	22.0 ± 7.5	30.2 ± 7.0
Peak VO <sub>2</sub> change in %	6.2	5.2

## P-84

**NT-proBNP increase at exercise in patients with univentricular heart after total cavopulmonary connection**

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**Background:** Increase of N-terminus pro B-type natriuretic peptide (NT-proBNP) during exercise was related to myocardial ischemia, myocardial dysfunction and inflammatory stress in

acquired heart disease. Its value in patients with univentricular heart after total cavopulmonary connection (TCPC) is unknown. **Patients and Methods:** 66 patients (19 female, age 8–52 years) with TCPC (lateral tunnel in 28 patients, extracardiac conduit in 38 patients) performed a symptom-limited cardiopulmonary exercise test on an upright bicycle ergometer. Venous NT-proBNP samples were drawn at rest and 2–3 min after peak exercise.

**Results:** Median NT-proBNP at rest was 82 ng/L (range 11–2554) with four patients above the upper reference limit of 480 ng/L. A higher NT-proBNP at rest was related to a worse aerobic capacity at the exercise test (ln NT-proBNP versus peakVO<sub>2</sub>, r = .333, p = .006).

The median increase of NT-proBNP at exercise was 6 ng/L (range 0–314 ng/L). All but one patient were below the published cut-off of 80 ng/L. In a multiple regression analysis, the absolute increase of NT-proBNP was mainly related to its resting value. The relative increase was solely related to a higher body mass (Δln NT-proBNP versus BM, r = .357, p = .003) and not related to any of the investigated functional parameter.

**Conclusions:** NT-proBNP at rest is usually not elevated in TCPC patients. If so, it is a valuable predictor of cardiac function. During exercise, there is only a minor increase in NT-proBNP. Its extent is considered normal in studies with adult patients and is not related to any of the investigated functional parameter. Maybe the filling restriction from the lungs prevents atrial and ventricular overload and BNP secretion in TCPC patients.

## P-85

**Pulmonary capillary haemangiomatosis: an unusual histological presentation of vascular lesion in lung biopsies from patients with congenital cardiac shunts and pulmonary hypertension**

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**Introduction:** The typical pulmonary arterial occlusive lesions occurring in patients with congenital cardiac shunts are characterized by medial layer hypertrophy and intimal proliferation. On the other hand, pulmonary microvasculopathy (former capillary hemangiomatosis) is a rare form of histological lesion of pulmonary hypertension (PH), usually described in the idiopathic or “primary” form of the disease, being sometimes associated to veno-occlusive lesions but not described in other etiologies. We report two cases of microvasculopathy in a large series of lung biopsies obtained from patients with congenital cardiac shunts.

**Patients and Methods:** Among 297 lung biopsies analyzed over an 11 year period, two from children with PH were found to have microvasculopathy. First patient is a 4 year-old girl with Down's syndrome, an 8 mm secundum atrial septal defect and stenosis of the right pulmonary artery. The other is a 2 year-old girl with a 13 mm perimembranous ventricular septal defect. The mean pulmonary arterial pressure was above 30 mmHg in both cases. No signs of left ventricular obstructive lesions were observed on echocardiography. Lung biopsies were performed to evaluate the severity of the arterial lesions and collected inflated in order to maintain the overall lung architecture. The first patient underwent closure of the atrial defect and enlargement of the right pulmonary branch and the second underwent pulmonary artery banding.

**Results:** Both biopsies showed severe hypertrophy of the walls of pre and intra-acinar arteries. Intimal proliferative lesions were present in pre and intra-acinar arteries from the first case and only

in pre-acinar arteries from the second one (Heath-Edwards grade II). Both cases showed thin-walled capillary vessels infiltrating the perivascular and peribronchial connective tissue and enlarging focally the alveolar walls. Hemosiderin laden macrophages were found in alveolar lumens. Such histological features are typical of pulmonary microvasculopathy. The cases reported here correspond to 1.5% of all lung biopsies in the series.

**Conclusion:** To the best of our knowledge, the occurrence of pulmonary microvasculopathy in association to the classical histological occlusive arterial lesions in congenital cardiac shunts has not been described. Its possible role in increasing the severity of pulmonary hypertension in this context and its pathogenesis need to be elucidated.

#### P-86

##### **Pre- and post-enteral feeding measurement of splanchnic/cerebral oxygenation ratio (SCOR) by near infra-red spectroscopy (NIRS) in neonates with congenital heart disease (CHD) and comparison with healthy neonates**

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An increased risk of necrotizing enterocolitis has been documented in neonates affected by CHD. However, the beneficial effect of early enteral feeding on the perinatal and surgical prognosis is well known.

Our study aims at evaluating the effect of enteral feeding on the splanchnic oxygenation in neonates with CHD, in comparison with healthy neonates.

The regional oxygen saturation index was measured by means of NIRS skin sensors positioned on the forehead and on the umbilical region of neonates with CHD and healthy neonates. The ratio between the splanchnic and cerebral oxygenation (SCOR) before and after enteral feeding was evaluated.

Twenty seven neonates affected by complex CHD were enrolled in the study (9 with LVOT obstruction, 6 with RVOT obstruction, 8 with TGA and 4 with HLHS) and compared with 18 healthy neonates. All the CHD neonates were in stable hemodynamic condition, on PGE1 at a mean dose of 35 ng/kg/min. The NIRS monitoring lasted 63,7 hours  $\pm$  32,6 in CHD patients and 5,4 hours  $\pm$  1,2 in controls. A mean of 4,7  $\pm$  1,7 meals were recorded in CHD patient, 2 meals in 8 controls and 1 in 10. CHD patients showed pre- and post-prandial SCOR significantly lower than controls: CHD SCOR pre 0.784  $\pm$  0.221, controls 1.019  $\pm$  0.134 (p 0.000), CHD SCOR post 0.793  $\pm$  0.217, controls 1.021  $\pm$  0.128 (p 0.000), without difference in pre- and post-prandial oxygenation. Among CHD patients, those with LVOT obstruction and HLHS had lower SCOR in comparison with TGA and RVOT obstruction patients, always without difference between pre and post prandial SCOR: LVOTO SCOR pre 0,76  $\pm$  0,19 post 0,75  $\pm$  0,18, HLHS SCOR pre 0,70  $\pm$  0,23 post 0,70  $\pm$  0,23; TGA SCOR pre 0,90  $\pm$  0,28 post 0,93  $\pm$  0,32; RVOTO SCOR pre 0,82  $\pm$  0,21 post 0,84  $\pm$  0,19.

**Conclusion:** In CHD patients, in stable condition, enteral oxygenation is significantly lower than in controls, mainly in patients with LVOTO and HLHS. Enteral feeding does not seem to modify the splanchnic oxygenation. According to these results the advantage of enteral nutrition on the surgical prognosis should overcome the risk of necrotizing enterocolitis.

#### P-87

##### **Blood pressure measurement procedure at an out-patient pediatric cardiology unit**

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Blood pressure measurements in children are recommended to be assessed as the mean of three recordings. In daily practice, however, blood pressure measurements are often assessed after one single recording. This procedure may be convenient for the child but may result in an unreliable estimate.

**Aim:** To compare blood pressure values obtained after one single recording with mean blood pressure values obtained after three consecutive recordings.

**Patients and Methods:** A total of 125 boys, median age 10.3 years (0.1–18.7) and 119 girls, aged 10.5 years (0.1–18.3) were recruited from February to December 2010, during their regular visit at a secondary out-patient pediatric cardiology unit. The children were divided into 5 groups: 1. Healthy, 2. Congenital heart defects, 3 Arrhythmias, 4. Cardiomyopathies and 5. Other diseases. Blood pressure was measured with the child in a sitting position in children over one year of age. Standard oscillometric equipment was used with three consecutive measurements completed during 10–15 minutes with the cuff on the right forearm. Results were given as one single value of the first recording and as the mean of three consecutive recordings for each child. All the children underwent ECG and echo-cardiography investigations which were normal in all the children in group 1 (Healthy).

**Results:** Systolic and diastolic blood pressure values decreased from the first to the third recording with a mean of 2.2 (–24 to 20) and 2.8 (–28 to 25) mmHg, respectively, in the total cohort of children (p < 0.0001). The first systolic and diastolic recordings were 1.0 (13 to –9) and 1.2 (17 to –10) mmHg higher than the mean values, respectively (p < 0.0001). The differences between the first and third recordings and one single recording and the mean of three, were the same in all the groups (p = 0.9 and p = 0.7, respectively). No correlations with age or length were observed.

**Conclusion:** Blood pressure values assessed as the mean of three recordings are slightly lower than those assessed after one single recording. The difference is trivial; however, indicating that one single recording of blood pressure is acceptable in children, provided the observed value is within the normal range.

#### P-88

##### **Long-term Outcome of Percutaneous Transluminal Coronary Rotablator Ablation for Ischemic Heart Disease Caused by Kawasaki Disease**

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**Objective:** The purpose of this study was to determine long-term outcome of percutaneous transluminal coronary rotablator ablation (PTCRA) in patients with ischemic heart disease caused by Kawasaki disease (KD).

**Methods:** Subjects were 15 patients (12 male and 3 female) with a history of KD who developed ischemic heart disease as a result of coronary aneurysms and were treated by PTCRA since 1994. From medical chart, patients' information concerning demographics, history of catheter and surgical interventions, and final outcome were collected. Based on these data, we calculated restenosis-free rate using Kaplan-Meier's analysis.



**Results:** Subject's age at onset was  $1.9 \pm 1.7$  years old and median observational period was 9.8 years (range 0.2–15.3 years). PTCRA was applied on #6 (n = 7), #1 (n = 5), #2 (n = 2), and #5 (n = 1) of coronary arteries using burr size of  $2.1 \pm 0.3$  (1.5–2.5) mm at median of 15.6 (4–23.9) years old,  $12.3 \pm 5.2$  years after KD. Coronary stenosis was alleviated from  $86 \pm 8.7$  to  $39 \pm 25\%$  stenosis with additional balloon dilation (13) or stent placement (2). In the follow-up period, 5 patients showed re-stenosis at 0.2, 2.7, 3.0, 10.2, 14.6 years after the initial PTCRA, giving re-stenosis free rate of 78% and 62% at 5 and 10 years after the initial PTCRA. Among these 5 patients, 4 patients underwent additional PTCRA with additional balloon dilation (3) and stent placement (1), but 1 patient who previously underwent coronary bypass graft surgery and had stenosis on #5 died at 2nd session because of acute myocardial infarction. The remaining 1 patient showed total occlusion of target vessel (#1), but was treated medically because of sufficient collateral communications. Of 15 patients, 14 patients survived with a median age of 23.8 years old.

**Conclusions:** Long-term outcome of PTCRA for ischemic heart disease caused by KD may not be acceptable and further refinement of the indication and technical innovation are necessary.

#### P-89

##### Withdrawn

#### P-90

##### Evaluation of the Cases with Silent Carditis

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**Introduction:** The first rheumatic fever attack may cause no clinical signs, but as a result of recurrent attacks may develop rheumatic heart disease. The aim of this study is to establish important of the silent carditis.

**Methods:** Between October 2005 and June 2010, 226 patients admitted to our section were included. The mean age was  $11,78 \pm 2,9$ , average follow-up period was  $30,2 \pm 10,98$  months. Valvar structure was detected by two dimensional and Doppler echocardiography. WHO echocardiographic criteria for making the diagnosis of silent carditis was accepted as the basis. Patients with rheumatic fever arthritis and chorea were defined as group 1 (78 patient); patient with chest pain, palpitation, syncope, murmur, athralgia and fulfilling the criteria of WHO were defined as group 2 (148 patient). All the patients were given the benzathine penicilin prophylaxy.

**Results:** All patients with Sydenham's chorea and 75% of patient with rheumatic fever arthritis showed signs of silent carditis in our study. 78 patients fulfilling the revised Jones criteria for rheumatic fever, 67 of the patients have had arthritis and 11 had chorea as major Jones criteria. Mitral insufficiency was mild in 89.4%, moderate in 10.6%, aortic insufficiency was mild in 88.8%, moderate in 11.2% of group 1 ; mitral insufficiency was mild in 89.2%, moderate in 10.8%, aortic insufficiency was mild in 88%, moderate in 12% of group 2. Mitral valve in 80.9%, aort valve in 3.8%, both valve in 15.3% of the group 1, mitral valve in 81.7%, aort valve in 3.7%, both valve in 14.6% of the group 2 was affected. Regression in mitral and aortic regurgitation were 62.6%–50% and 64.2%–52% in group 1 and 2 respectively. Mitral and aortic regurgitation length and velocity was significantly decreased in both group during follow up (  $p < 0,001$ ). No patient had progression of valvular lesions. There was no difference between two groups in terms of improvement of valvular lesions.

**Conclusion:** Subclinical lesions aren't evanescent and could be detected by Doppler echocardiography. We suggest that

echocardiographic findings should be accepted as a major criterion for the diagnosis of rheumatic fever.

#### P-91

##### Pathognomonic ECG in Neonates with Enterovirus Myocarditis

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**Objective:** To assess whether there is a distinctive ECG pattern in neonates who developed enterovirus (EV) myocarditis within the first weeks of life.

**Design:** Electrocardiogram (ECG) findings, clinical presentation, echocardiography data, and the outcome of infants with EV myocarditis were retrospectively analysed and compared with the literature.

**Results:** From 1994 to 2009 eighth neonates presented with cardiac failure within 17 days after birth requiring respiratory and circulatory support. Cocksackie virus B could be detected as the source of the myocarditis in all. Two infants died 10 and 39 days after diagnosis.

ECG performed at admittance showed a complete loss of the R-wave and a deep Q wave in I, II and the left precordial leads (black arrows in figure) in all patients. These ECG changes resolved within months in the survivors. Echocardiography showed left ventricular dilatation, severe systolic dysfunction and hyperechogenic non contractile areas in all; analogous to myocardial infarction in adults. In all survivors aneurysm formation in the left ventricular wall was found weeks to months later. The survivors developed long-term cardiac sequelae requiring medication. Only one similar ECG in a neonate with EV myocarditis was published in the literature without comparing it with the whole cohort (Inwald D et al, Arch Dis Child Fetal Neonatal Ed. 2004;89:F461-2)

**Conclusion:** Complete loss of R-wave and pathologic Q waves seem almost pathognomonic for EV myocarditis in neonates which predominantly affects the left ventricle. This ECG pattern is not known as typical ECG pattern in acute myocarditis. It is comparable with ECG after myocardial infarction, but unlike patients with myocardial infarction this ECG pattern returns to normal in surviving patients. This phenomenon is not described earlier in the literature. ECG should be used as a simple diagnostic tool after admittance of a neonate with the history of suspected neonatal myocarditis. It does not replace virus diagnostic or echocardiography, but confirms and completes its diagnosis.

#### P-92

##### Clinicopathological investigation on pediatric cardiomyopathy with special reference to ongoing myocardial damages using pathophysiological parameters

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**Purpose:** Clinical significance of ongoing myocardial damage in pediatric cardiomyopathy has not been well defined. Furthermore, it is not so easy to estimate the severity of myocardial damages only from clinical symptoms and signs. To clarify the possible

prognostic impact of ongoing myocardial damages, we investigated pathophysiological parameters, such as symptoms and signs related to HCM, dysrhythmias and ST-T and changes by Holter ECG and exercise ECG, electrophysiological study, biochemical markers and endomyocardial biopsy (EMB).

**Patients and method:** They included 18 hypertrophic cardiomyopathy (HCM); with pressure gradient 7, without 11, aged from 2.5 to 18 years, 7 patients with dilated cardiomyopathy (DCM); aged from 1 to 15 years. Male to female was 13 to 12. Selected biochemical markers in this study were high-sensitive C-reactive protein (hsCRP), myoglobin, Creatin Kinase MB (CK-MB), troponin T (TnT), heart-type fatty acid binding protein (H-FABP), ANP and BNP. Histopathology was evaluated with histomorphometric method. The fraction of myocardial volume occupied by fibrillar collagen (% fibrosis) and diameter of myocytes were detected by quantitative morphometry with an automated image analysis system.

**Results:** Death or resuscitated sudden death occurred in 7 patients. Hypotensive response on rapid atrial pacing on EPS and exercise test was in 4. Pacemaker implantation was required in 2. Myocardial changes on EMB showed degeneration, interstitial fibrosis and disarray of myofibrils, inflammatory cell infiltration and microangiopathy. % fibrosis showed higher in cardiac death patients. Prevalence of abnormal biochemical markers was found in 35% of H-FABP, 35% of myoglobin, 32% of CK and 60% of ANP and BNP.

**Conclusions:** Many cases with cardiomyopathy showed raised concentration of biochemical markers. Although the correlation between H-FABP concentration and heart failure severity was reported in a previous study, in this study clinical severity did not reveal statistic correlation with biochemical markers and histopathological severity obtained by endomyocardial biopsy. Nevertheless, H-FABP and other biochemical markers in cardiomyopathy might be one of the plausible predictors for the ongoing myocardial damage and ongoing ischemia. An aggressive diagnostic evaluation to detect ischemia and the use of a multifaceted treatment approach to prevent ischemia and sudden death may be beneficial.

### P-93

#### Levosimendan on severe low cardiac output in paediatric patients

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**Introduction:** Levosimendan is a myocyte calcium-channel sensitizer and it is an optional drug to treat severe low cardiac output (SLCO).

**Objective:** To report the outcomes in patients (p) with SLCO receiving intravenous levosimendan as part of SLCO treatment strategy.

**Methods:** Descriptive-retrospective and observational study of 25 patients with diagnosis of SLCO requiring IV levosimendan. Cardiogenic shock was diagnosed based on clinical finding, ECG, 2D-echocardiography. Central venous oxygen saturation (ScvO<sub>2</sub>), arterial oxygen saturation (SaO<sub>2</sub>), arterial-venous difference saturation (AV dif sat O<sub>2</sub>), systemic oxygen extraction index (SO<sub>2</sub> ext index) were measured immediately before and 24 hs after intravenous levosimendan administration (loading dose 6 µg/kg/m – maintenance dose 0,1 µg/kg/m during 24 hs)

**Results:** 25 p were enrolled between January 2006 to December 2009. Mean age was 3.0 years (1 m–16 y), 11 p were female and the mean weight was 12.0 kg (3.1–55). Diagnosis: 14 were post-op cardiovascular surgical patients (4 p TOF, 3 p AVC, 3 p d-TGA, 1 p TOF + AVC, 1 p modified Fontán-Kreutzer

procedure, 1 p type B IAA, and 1 p TAPVD). 8 p had been diagnosed with acute myocarditis (5 p post scorpion poisoning, 1 p mycoplasma +, 1 p adenovirus+ and 1 p without confirmed etiology), 2 p had dilated cardiomyopathy (1 p with end-stage myocarditis and 1 p with non compaction LV) and 1 p with long QT syndrome with syncope.

After levosimendan administration, all p showed HR decreased from 175 beats/min (r: 155–210) to 150 beats/min (r: 123–170), mean BP increased from 48 mm Hg (r: 35–70) to 60 mm Hg (50–77), LV shortening fraction improved from 11% (r: non-detectable–22%) to 23% (14–25), SvcO<sub>2</sub> increased from 59% (45–60) to 71% (58–79). All this variables showed a significant statistical difference with p < 0.001. AV dif sat O<sub>2</sub> decreased from 42% to 31% (p: 0,003) and SO<sub>2</sub> ext index decreased 0,42 to 0,31 (p < 0,003). 4/25 p (16%) died (2 p belonging to the post-op group, 1 p post scorpion poisoning and 1 p with long QT syndrome).

**Conclusion:** In our experience, levosimendan administration in pediatric patients suffering SLCO is a safe and effective treatment option with favorable outcomes.

### P-94

#### Non-invasive monitoring of microcirculation in paediatric patients suffering from septic shock

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**Introduction:** Oxygen related variables and diastolic blood pressure (DBP) allow to estimate the systemic vascular resistance (SVR). Low central venous oxygen saturation (ScvO<sub>2</sub>), and an increase in the arterial-venous difference saturation (AV dif sat O<sub>2</sub>) as well as in the systemic oxygen extraction index (SO<sub>2</sub> ext index) indicate ↑SVR. An increase in ScvO<sub>2</sub> and a decrease in AV dif sat O<sub>2</sub> and DBP, indicate ↓SVR.

**Objectives:** To identify hemodynamic trends and assess pharmacological strategies in pediatric patients with septic shock.

**Methods:** Retrospective – descriptive and observational study. Between January 2008 and December 2009, 35 patients (p) were enrolled (p) in the present study with diagnosis of septic shock (23 males), with an average age of 14 months (range: 1 month to a 15 years).

**Results:** Group A (non-hyperdynamic shock): 24 p, 17 had myocardial dysfunction and 3 had hypovolemic state. 10 p showed isolated hypovolemia. Systolic blood pressure (SBP) < P5 and DBP > P50. ScvO<sub>2</sub>: 53,4%; AV dif sat O<sub>2</sub>: 40,9%; SO<sub>2</sub> ext index 0,43, AV dif CO<sub>2</sub>: 10,7 mm Hg and ↑lactate.

Fluid administration (10–60 ml/kg) and vasodilators (dobutamine: 6 p, dopamine + milrinone: 8 p and epinephrine: 4 p) were started. 8 p shifted to group B, 3 p improved with norepinephrine, 3 p with methylene-blue and 2 p with terlipresine.

Group B (hyperdynamic shock): 11 p, 5 p had myocardial dysfunction and 3 p with hypovolemic state. Normal SBP and DBP < P5; ScvO<sub>2</sub>: 75,3%; AV dif sat O<sub>2</sub>: 19,9%; SO<sub>2</sub> ext index: 0,21; AV dif CO<sub>2</sub>: 9,1 mm Hg and ↑lactate. All p received vasopressors (dopamine + norepinephrine). 2 p showed refractory vasoplegic septic improving with methylene-blue.

Overall mortality was 8 p (28%).

Variable	Group A	Group B	p value
ScvO <sub>2</sub>	53,4 ± 5,25	75,3 ± 1,25	0,00001
AV dif sat O <sub>2</sub>	40,9 ± 3,02	19,9 ± 1,53	0,00001

**Conclusion:**

1. ScvO<sub>2</sub> and oxygen derives variables allow to assess the microcirculation.
2. Detection of hemodynamic trends lead to a rational drug support and fluid administration.
3. Methylene-blue administration emerges as an attractive alternative in p with refractory vasoplegic shock.

**P-95****Recurrent pericarditis as a sole manifestation of Familial mediterranean fever**

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**Introduction:** Familial Mediterranean fever (FMF) is characterized by recurrent episodes of inflammation and serositis including fever, peritonitis, synovitis, pleuritis and less frequently pericarditis and meningitis. Isolated pericarditis cases are rare.

On the other hand, the most common causes of recurrent pericarditis are: virus, idiopathic and post-pericardiectomy.

**Case reports:** We present three case reports of Spanish adolescents from 11, 14 and 16 years old who presented with clinical recurrent episodes of pericarditis. No history of surgery or other tissue inflammation was reported. Two of them responded to treatment with colchicine, reducing pericarditis episodes, while another one did not require base treatment. Genetic analysis to FMF diagnosis resulted in different heterozygous mutations in MEFV gene, which have been associated with FMF.

**Conclusion:** Faced with recurrent pericarditis unresponsive to standard treatment with NSAIDs and steroids, cardiologist should always bear in mind to exclude the FMF diagnosis.

It is remarkable that these patients with recurrent pericarditis as a sole manifestation of FMF are heterozygous, so they would have to be considered as carriers and would have no risk of developing the disorders. However it is not so and could be explained by the presence of unknown allelic mutations not tested.

**P-96****Relationship of TNF- $\alpha$ -308, IL-10-1082 Gene Polymorphisms with the Severity and Susceptibility of Rheumatic Heart Disease**

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**Introduction:** Acute rheumatic fever is an inflammatory disease developing after upper respiratory tract infection with group A streptococci and its most important complication is rheumatic heart disease. Recent studies emphasize the importance of IL-10 and TNF- $\alpha$  gene polymorphism in the pathogenesis. There are limited numbers of studies reporting TNF- $\alpha$ -308 and IL-10-1082 gene polymorphism may induce susceptibility to rheumatic heart disease. Gene polymorphisms change depending on race, and in Turkey there is no study on IL-10-1082 gene polymorphism in the patients with ARA. However, there are only two studies on TNF- $\alpha$ -308 of which results were conflicting. The aim of our study is to determine the frequency of IL-10-1082 A/G and TNF- $\alpha$ -308 G/A gene polymorphism in Turkish population and to investigate the relationship between these polymorphisms and rheumatic heart disease.

**Methods:** In this case-control study, the relationship between G/A polymorphisms in TNF- $\alpha$ -308 gene, A/G polymorphism in IL-10-1082 gene and rheumatic heart disease and valvular

involvement. A total 57 patients with rheumatic heart disease and 99 healthy controls were included.

**Results:** The rate of TNF- $\alpha$ -308 gene polymorphism was %3,1 in healthy subjects and this polymorphism was not observed in patients with rheumatic heart disease. In healthy subjects, the frequency of IL-10-1082 gene polymorphism was higher than the patients with rheumatic heart disease. There was no relation between TNF- $\alpha$ -308 genotype and allele distribution with valvular involvement ( $p > 0.05$ ). IL-10-1082 G/G and A/G genotypes were seen more frequent in patients with multiple valvular disease but there was no statistical significance ( $p > 0.05$ ). **Conclusion:** As a result, there was no relationship between TNF- $\alpha$ -308, IL-10-1082 gene polymorphisms and rheumatic heart disease or valvular involvement in the study population ( $p > 0.05$ ). TNF- $\alpha$ -308 polymorphisms are silent and may become important only with some certain HLA alleles. Studies checking both cytokine polymorphism and HLA alleles are needed.

**P-97****Neonatal Heart Failure in Lagos University Teaching Hospital Nigeria**

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University College Hospital, Ibadan, Nigeria (2)

**Background:** Heart failure in the neonate is an important cause of morbidity and mortality in the developing world. The aim of this study was to determine the prevalence, specific aetiology, outcome and to highlight some short term factors affecting outcome among cases of heart failure admitted into the neonatal wards of Lagos University Teaching Hospital (LUTH), Lagos Nigeria.

**Methods:** Forty-four consecutive cases of heart failure admitted into the neonatal wards of Lagos University Teaching Hospital, Lagos with diagnosis of heart failure over one year period was studied prospectively. Diagnosis of heart failure was based on the presence of at least three of the four cardinal signs of heart failure: tachycardia, tachypnoea, tender hepatomegaly and Cardiomegaly. All cases were followed up daily till a definite outcome was determined.

**Results:** Neonatal heart failure constituted 2.7% of the total neonatal admissions during the study period. The predominant underlying causes were severe anaemia (34.1%), neonatal sepsis (22.7%), and congenital heart disease (20.5%). Other causes were perinatal asphyxia (13.6%), and acute respiratory infections (9.1%). There was a case-fatality rate of 27% among the study population. Factors influencing outcome were age below 24 hours or above seven days, neonates delivered outside LUTH (outborns), and late presentation to hospital after seven days of illness.

**Conclusion:** Heart failure in Nigerian neonates though mostly due to preventable causes, are associated with an unacceptably high mortality.

**P-98****Left ventricular systolic and diastolic function in children with overweight and obesity**

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**Objective:** Many studies have proven the relation between obesity and heart failure. The purpose of our study was to identify the influence of the severity of obesity in children on structural and functional changes of the left ventricle and parameters of the systolic and diastolic function.

**Design:** Prospective controlled study. 21 children with overweight and obesity and 23 healthy controls.

**Subjects:** Following parameters have been evaluated: nutritional status, intermittent and continuous ambulatory blood pressure monitoring, echocardiographic examination including aortal root diameter, left atrial diameter, interventricular septum thickness, left ventricular end-diastolic diameter, left ventricular end-systolic diameter, left ventricular posterior wall thickness, left ventricle mass, relative wall thickness, left ventricular mass/height index and ejection fraction.

**Results:** In overweight and obese children, systolic blood pressure, diastolic blood pressure, average day-time SBP were higher than in controls. Left ventricular size and function parameters were also increased in overweight children. There was diastolic dysfunction with preserved systolic function in study group.

**Conclusions:** In our study we have found, that in obese and overweight children signs of early myocardial damage, both structural and functional, are detectable mainly as the alteration of the left ventricular diastolic function, despite preserved global systolic function. These changes seems to be intensified by the severity of obesity.

### P-99

#### Impact of chest X-ray before discharge in children after cardiac surgery – prospective evaluation

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**Introduction:** Chest X-rays are performed routinely before discharge after cardiac surgery in many paediatric cardiac units. These radiographs contribute to radiation exposure, therefore indications for chest X-rays should be restrictive.

**Objective:** To evaluate the diagnostic impact of routine chest X-rays before discharge in children undergoing open heart surgery and to analyze certain risk factors predicting pathologic findings.

**Methods:** In a prospective single centre observational clinical study 128 consecutive children undergoing heart surgery (mean age 28 months, range 0–17.9 years, 69 male) received a biplane chest X-ray 13 days (mean) after operation, before planned discharge. Pathologic findings in chest X-rays were defined as infiltrate, atelectasis, pleural effusion, pneumothorax or signs of fluid overload/pulmonary hypercirculation. Their therapeutic consequences were documented. 109 asymptomatic children were included in the final analysis. Risk factors such as age, corrective versus palliative surgery, reoperation, sternotomy vs. lateral thoracotomy and pulmonary complications (i.e. pulmonary infection, pleural effusion, atelectasis, pneumothorax) during postoperative ICU (Intensive Care Unit) stay were analysed.

**Results:** In only 5.5% (6/109) of these asymptomatic patients pathologic findings in routine chest X-ray before discharge were found – pleural effusion (n = 1), atelectasis (n = 1), pneumothorax (n = 1), signs of fluid overload (n = 3). In only three of these cases (50%) subsequent non-invasive medical intervention (increasing diuretics) was needed. Five of these six patients have had complications during ICU stay. Risk factor analysis revealed only pulmonary complications during ICU stay to be significantly associated (p = 0.036) with pathologic X-ray findings before discharge.

**Conclusions:** Routine chest X-rays before discharge can be omitted in asymptomatic children after cardiac surgery with an uneventful and straightforward perioperative course. Chest X-rays before discharge are warrantable if pulmonary complications during ICU stay did occur as this is a risk factor for pathologic findings.

### P-100

#### Impact of acute cardiac transfers conducted by the West Midlands Neonatal Transfer Service

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**Introduction:** Congenital heart disease is the commonest group of congenital malformations and accounts for approximately 10% of infant deaths.<sup>1</sup> These infants need to be transferred to centres with expertise in paediatric cardiology and account for a significant proportion of transfers conducted by neonatal transfer teams. The West Midlands Neonatal Transfer Service (WMNTS) was established in 2007 and is primarily led by Advanced Neonatal Nurse Practitioners (ANNPs). It provides transport within neonatal networks comprising of 16 neonatal units within the West Midlands region. **Objective:** To review the impact of cardiac transfers conducted by the WMNTS for congenital heart disease.

**Methods:** Retrospective review of all acute cardiac transfers conducted by WMNTS from Jan 2007 to Aug 2009.

**Results:** A total of 236 transfers were conducted during the study period. 188 babies were on a prostin infusion of which 45 babies were ventilated. All babies on a high dose prostin infusion were transferred ventilated. 184 were transferred to the cardiology wards, while 52 were transferred to PICU. 225 babies were transferred to the regional tertiary children's hospital. There were no reported clinical incidents.

	2007	2008	2009
Total number of cardiac/total annual transfers	77/1115	79/1251	80/ 1328
Median gestational age in weeks (range)	40 (31–42)	39 (31–42)	39 (32–43)
Median birth weight in grams (range)	3090 (1096–4400)	2960 (1020–4890)	3100 (1064–4536)
Male : Female ratio	36: 41	42: 37	42: 38
Receiving prostin	59	55	74
Prostin dose in nanograms/kg/min (range)	5 (5–100)	5 (5–100)	5 (5–100)
Ventilated	17	13	15
ANNP: Registrar	71: 1	59:13	50:24

**Conclusions:** Acute cardiac transfers comprise approximately 6% of the transfers conducted by WMNTS. 95% of the babies remained within the region. All transfers were completed safely. ANNPs are capable of transferring these sick babies safely.

Reference:

1. Abu-Harb M. et al. Arch Dis Child 1994; 71: 3–7.

### P-101

#### Takayasu arteritis in children

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**Objective:** Show our experience in clinic manifestations and evolution in infants with Takayasu arteritis.

**Material and Methods:** We made a retrospective, transverse and observational study, where we analyzed the frequency, clinical and laboratory manifestations treatment and clinical evolution in patients with Takayasu arteritis in the last 28 years.

**Results:** We obtained 30 patients with gender relation 1:1, with age from 4 months to 16 years old, with mean age 11 years old. The main clinical manifestations were cardiovascular, 25 patients with systemic arterial hypertension and 10 with different pulses and neurologic clinical manifestations like second in frequency. In Zapata classification of arterial affected areas, we found it 14 patients in V type and 7 in III type.



The laboratory exams more frequent were: 12 with proteinuria, positive exam PPD in 11 and anemia in 9 patients. Thirteen patients with left ventricular hypertrophy.

Treatments in the patients were: steroids in 26, antihypertensives in 25, ciclofosfamide in 15.

The monitoring time was of 11 years. Twenty four patients had a good clinic evolution, 6 of these died (20%).

**Conclusions:** We suggest that the clinic manifestations of Takayasu arteritis were different in infants than in adults.

All the infants with systemic arterial hypertension, should be studied for Takayasu arteritis diagnosis.

The prognosis of Takayasu arteritis in infants has been better in the last years than the last century.

## P-102

### Enhanced aortic pressure wave reflection in patients after successful repair of aortic arch in children

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**Introduction:** Despite an apparently successful surgical repair of aortic coarctation, subsequent cardiovascular complications (hypertension, ischemic heart diseases, stroke etc.) have sometimes been encountered. Increased aortic pressure wave reflection is one of the risk factors for developing such cardiovascular diseases, and the enhanced pressure wave reflection has been reported in patients after an aortic arch repair. The purpose of this study is to clarify whether the enhanced pressure wave reflection becomes cardiac load in patients with aortic coarctation after successful aortic arch repair.

**Methods:** This study enrolled 25 patients aged 1–25 year ( $9.8 \pm 5.6$ ) with aortic coarctation (19) or interrupted aortic arch (6) after a successful aortic arch repair (i.e. no pressure gradient in aortic arch). The methods of aortic arch repair were extended to end-to-end anastomosis in 15, subclavian flap in 7, Blalock–Park operation in 1, patch angioplasty in 1 and a graft interposition in 1. The period after aortic arch repair was  $8.5 \pm 4.2$  years. None of them were taking any medication and none were diagnosed to be suffering from Turner syndrome. The aortic pressure waveform was recorded using a pressure sensor mounted catheter, and the augmentation index was thus calculated. The relationship between the increment of augmentation index and left ventricular mass, which was calculated by using echocardiography, was examined.

**Results:** The increment of augmentation index was  $27.7 \pm 16.7\%$ . The percent of left ventricular mass was  $124.3 \pm 25.4\%$  and left ventricular mass index was  $85.7 \pm 18.9 \text{ g/m}^2$ . There was positive correlation between the increment of augmentation index and the percent left ventricular mass ( $p = 0.026$ ), and left ventricular mass index ( $p = 0.025$ ).

**Conclusions:** In patients after a successful aortic arch repair, the pressure wave reflection increases, and it is one of the causes of left ventricular hypertrophy of them. The enhanced pressure wave reflection may lead to future cardiovascular disease in patients after repair of aortic arch.

## P-103

### Low agreement between cardiologists diagnosing left ventricular hypertrophy in children with End Stage Renal Disease

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**Introduction:** Early detection and treatment of left ventricular hypertrophy (LVH) is essential in children with end stage renal disease (ESRD) to prevent cardiac mortality. To date, LVH diagnosed by echocardiography is believed to be the most appropriate surrogate marker for cardiac disease in these children. However, in the multicentre RICH-Q project, the incidence of LVH appeared to differ considerably between centres. Therefore, we assessed the intra and inter observer reproducibility of the measurement of the diastolic inter ventricular septum (IVSd) and posterior wall (LVPWd) thickness.

**Methods:** Digital images of the echocardiograms of 92 children with ESRD aged 0–18 years from 4 different centres were analyzed offline by three independent observers. Kappa was calculated to assess inter observer consistency. The measurement errors are expressed as Smallest Detectable Change (SDC).

To exclude within-patient variability two observers also assessed one selected image from twenty echocardiographies, representing the entire range of the patient population.

**Results:** Kappa between the 4 observers ranged from 0.1–0.4, which is considered low. The intra observer SDC ranged from 1.6–1.7 mm and from 1.3–2.6 mm for IVSd and LVPWd, respectively. The inter observer SDC were 2.4 mm and 2.6 mm for IVSd and LVPWd, respectively. For selected images the intra observer SDC ranged from 1.0–1.3 mm and from 0.7 to 2.1 mm, the inter observer SDC were 2.1 and 2.3 mm for IVSd and LVPWd, respectively.

**Conclusions:** Agreement between different observers diagnosing LVH using conventional echocardiography is low. In individual children changes in diastolic wall thickness smaller than 1.6 mm cannot be distinguished from measurement error, even when measured by the same observer. This limits the use of echocardiography to detect changes in IVSd or LVPWd in children with ESRD, which may have important clinical consequences.

## P-104

### Echo Doppler Assessment of Vascular Function in Post-operative Congenital Heart Disease

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**Background:** Most children with congenital heart disease (CHD) now survive until adulthood with a good quality of life. In some forms of CHD the aorta may be large and abnormal which has implications for future cardiovascular risk. Therefore, we sought to assess the biophysical properties of the aorta of children with: 1) tetralogy of Fallot (TOF); 2) coarctation of the aorta (COA) and 3) transposition of great arteries (TGA).

**Methods & Results:** Forty children with CHD. (17 TOF, 16 COA and 7 TGA) were compared with 55 matched control subjects (CTRL). The aortic diameters and pulse wave transit time around the aortic arch were measured with echo-Doppler. Pulse wave velocity (PWV), aortic input impedance ( $Z_i$ ), characteristic impedance ( $Z_c$ ), arterial pressure-strain elastic modulus (Ep) and

arterial wall stiffness index ( $\beta$ -index) were calculated. The results are presented in Table 1. There was no significant difference within the CHD group.

**Conclusion:** Children with certain forms of CHD have impaired biophysical properties of the aorta, with increased PWV, impedance and stiffness. This predisposes them to early-onset cardiovascular events such as elevated blood pressure and ischemic events. Since these measurements are easily obtainable by routine ultrasound, they should become part of the regular follow-up of children with certain forms of CHD.

Table 1. Demographics data and biophysical properties of the aorta.

	CTRL group (n = 55)	CHD group (n = 40)	P value
Age [years]	13.8 +/- 4	14.5 +/- 2.6	0.36
PWV [cm/s]	351.4 +/- 49.9	540.9 +/- 174.6	<0.001
Zi [dyn · s · cm <sup>-5</sup> ]	140.5 +/- 27	134.4 +/- 60.9	0.21
Zc [dyn · s · cm <sup>-5</sup> ]	132.9 +/- 31.4	193.4 +/- 89.6	0.003
Ep [mm Hg]	247.9 +/- 52.1	310.6 +/- 127.6	<0.001
$\beta$ -index	3.0 +/- 0.6	3.9 +/- 1.5	<0.001

### P-105

#### Fewer Electrocardiogram Abnormalities in Duchene Muscular Dystrophy Patients Treated With Bisphosphonates

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**Introduction:** Cardiac involvement in Duchene muscular dystrophy (DMD) is well described with varying onset and severity of electrocardiogram (ECG) and echocardiogram changes. A regional cohort of patients with DMD receiving steroid therapy and bisphosphonates managed in a single centre showed improved survival (Pediatrics, in press). This study describes the possible effect of therapy on ECG abnormalities.

**Methods:** The cardiology records of a previously described regional cohort of patients with confirmed DMD who were born from 1963 to 2006 and who had received at least one year of steroid therapy were reviewed. Available ECGs were reviewed by a cardiologist (KKW). The following measurements were abstracted: R V1, S V1, R V6, S V6, Q in V6, and Q in III and compared to available age appropriate standards. Given the small sample size, tabular analysis used a two tailed Fisher's exact test.

**Results:** Forty-four boys with DMD from this cohort were exposed to continuous steroid use. Bisphosphonate therapy had been initiated for 16 patients (36%) at a median age of 12.5 years (range: 7–23 years). ECGs were available for review for 26 (59%). Single ECGs were available for 69% of the sample and serial ECGs for the remainder. One ECG measurement appeared to be associated with bisphosphonate use, a normal R V1 ( $p = 0.10$ ). When only children in their second decade were reviewed, the relationship between a normal R V1 and bisphosphonate use appeared stronger (OR 15,  $p = 0.04$ ) and for those with serial ECG data, all who had abnormal R V1 measurements, 3 receiving bisphosphonate therapy improved to normal, with 5 not receiving bisphosphonate therapy remained abnormal ( $p = 0.02$ ).

**Conclusions:** The treatment of patients with Duchene muscular dystrophy with steroid and bisphosphonate may be associated

with improved and/or fewer patients with ECG criteria for right ventricular hypertrophy.

### P-106

#### Non-compacted cardiomyopathy: clinical characteristics, evolution and prognostic data in childhood. Results of a multicentre study

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**Introduction:** Noncompaction of the ventricular myocardium (NCVM) is a rare congenital heart disease (CHD). Heightened awareness has resulted in an increased detection of the morphological features of NCVM in routine clinical practice.

**Methods:** Multicentre study (6 Catalan hospitals) including paediatric patients affected by NCVM according to echocardiographic criteria of Chin and Jenni. Clinical features, echocardiographic, MRI, complications and treatments are reviewed. Descriptive data and statistical analysis is provided.

**Results:** A total of 29 patients were included, 15 female and 14 male, the median age at diagnosis was 5 years and 7 months (range from birth to 17 years). In four cases (14%) cardiomyopathy was detected in utero. Sixteen patients (55%) presented as an isolated lesion, 8 (27.5%) had a ventricular septal defect associated one of them with aortic coarctation, 3 (10%) had an inborn error of metabolism, 1 (3.5%) had Juvenile Idiopathic Arthritis and 1 (3.5%) has a syndrome being studied. The location of the trabeculae has been predominantly at the apex, but also affected the left ventricle free wall in 11 patients (40%) and right ventricle in 2 (7%). MRI findings provided no further information. No complications have occurred in 12 patients (41%), cardiac failure 12 patients (41%), implantable cardioverter defibrillator was placed for ventricular arrhythmias in 2 patients (7%), stroke 1 patient (3.5%) and death 2 patients (7%), both of them less than 6 month of age ( $p < 0.05$ ). Median follow up has been 12 month (range from 2 months to 8 years). Current treatment includes carvedilol, ACEI's and ASA and one patient is waiting for cardiac transplantation.

**Conclusions:** Early onset of symptoms is related with poor prognosis. The clinical and prognostic heterogeneity described supports the theory that NCVM is not a disease but a morphologic feature.

### P-107

#### Standard values for the 6-minute walking distance in healthy children and adolescents from different nations – which one to rely on?

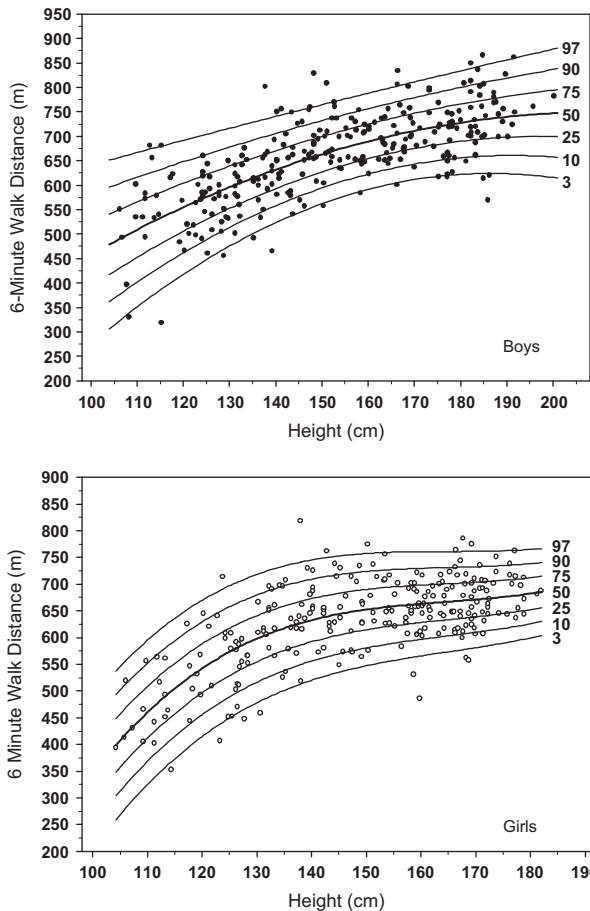
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**Introduction:** The 6-minute walk test (6-MWT) represents the most practicable and reliable test method to evaluate functional capacity on a submaximum level.

Recently reference values for the 6-minute walking distance (6-MWD) for healthy children and adolescents from four different countries have been published. Which one is better suited, given the methodological differences and different results?

**Methods:** Comparison of the results from 4 published studies on reference values for the 6-MWD in children and adolescents (two european, one chinese and one american), and interpretation of potentially confounding factors.



**Results:** 6-MWD of 3 study groups (austrian, american and chinese children and adolescents) correlated quite well despite different, albeit standardized, methodological approaches. Values published by the British study group were substantially lower than those of the other groups. Potentially confounding factors are age, height, ethnicity of the studied population. Because of these imponderables we propose sample-based centile curves for boys and girls for the standard values of the 6-MWD in children and adolescents of caucasian origin. Height was used for the construction of our standard reference curves because of its most discriminative properties of all anthropometric factors.

**Conclusions:** Results of the 6-MWT in children differ between different populations. Growth and methodological issues have to be taken in account when conducting comparable studies.

#### P-108

##### **Electrocardiographic Findings at Initial Diagnosis in Children with Isolated Left Ventricular Noncompaction**

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**Background:** The aim of this study was to comprehensively evaluate electrocardiographic (ECG) findings of IVNC patients

at initial diagnosis and to explore the correlation between them and the clinical, echocardiographic and magnetic resonance imaging (MRI) findings.

**Methods:** 23 patients diagnosed with IVNC by echocardiography and cardiac MRI between January 2006 and June 2010 were enrolled in this study. The patients were examined with standard ECG and 24 hour Holter ECG. For comparison purpose, ECGs of 50 healthy children of similar ages and demographic characteristics were taken.

**Results:** In 87% of patients ECG abnormalities were found. The most frequently seen ECG findings were left ventricular hypertrophy, ST segment depression and negative T wave related to abnormal repolarization particularly in D II, D III and V4-6 leads, as well as prolonged PR and QTc intervals. No ECG features or patterns were found that were specific to the disease. In contrast to adult patients, while no intraventricular conduction defects (particularly in the left bundle branch) were found in any of our patients, 13% had considerable bradycardia and one required a pacemaker. The Holter ECG recordings showed supraventricular tachycardia attacks in two patients and a short ventricular tachycardia attack in one. Patients whose echocardiograms and MRI showed left ventricular systolic dysfunction and left ventricular dilatation had signs of left ventricular hypertrophy and repolarization abnormality on their ECGs, but there was no significant difference in PR, QRS, and QTc intervals.

**Conclusion:** Regardless of how frequently left ventricular hypertrophy and repolarization abnormalities are found on IVNC patients' initial ECGs, we think that they are not unique to the disease, but are related to the severity of the cardiomyopathy.

#### P-109

##### **Left ventricular noncompaction in children and adolescents: clinical features, treatment and follow-up**

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**Objective:** Left ventricular noncompaction (LVNC) is a specific cardiomyopathy that occurs following a disruption of endomyocardial morphogenesis. This study presents clinical findings, diagnostic features, treatment and follow-up of pediatric patients diagnosed with LVNC.

**Methods:** Patients with LVNC who were followed from January 2006 to March 2010 were included in this study. Diagnosis was made with the use of characteristic findings of magnetic resonance imaging and echocardiography. Holter electrocardiography and metabolic screening tests were also performed in all patients.

**Results:** A total of 24 patients were studied (18 male, 6 female). Patient age at diagnosis was  $50 \pm 60$  months (8 days to 15 years). Average follow-up period was  $22 \pm 12$  months (4 months to 4 years). Findings at diagnosis were as follows: 8 (33%) patients had heart failure, five (20%) had rhythm abnormalities, five (20%) had cardiomegaly, two had murmurs, two – cyanosis, and two presented with fatigue. Ten (41%) patients had been followed previously with other diagnoses. In 21 (87.5%) patients, electrocardiographic abnormalities were noted, especially left ventricular hypertrophy and ST-T changes. Patients had an average ejection fraction of 46% (18–73%) and three of them had additional congenital heart disease (patent ductus arteriosus,

aortopulmonary window and complex cyanotic heart disease). Scanning for metabolic diseases revealed fatty acid oxidation disorder in one patient, and mitochondrial disease in another. During follow-up, a permanent pacemaker was implanted in a patient with severe bradycardia and ventricular dysfunction, and 3 patients died.

**Conclusion:** LVNC can be diagnosed at any age from newborn to adolescent and has a variable clinical course. Closer study of patients with cardiomegaly and heart failure can reduce delays in diagnosis of LVNC.

#### P-110

##### **Outcomes post Kawashima procedure and hepatic vein redirection for single ventricle palliation in left atrial isomerism with interrupted inferior vena cava**

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**Objectives:** Patients with left atrial isomerism (LAI) and interrupted inferior vena cava (IVC) palliated with a superior cavopulmonary connection or Kawashima procedure (KP) have a high incidence of developing pulmonary arteriovenous malformations (PAVMs). We aimed to assess the clinical outcomes of patients with LAI following a KP. The main end points were death, requirement for hepatic vein redirection (HVR) and its impact on oxygen saturations. We also sought to identify predictive factors for the necessity, or timing of HVR.

**Methods:** Retrospective data review of 24 patients with a diagnosis of LAI, interruption of the IVC and single ventricle physiology managed with a KP at a single centre between January 1990 and March 2010.

**Results:** 21 patients had a KP, with 12 subsequently undergoing a HVR. There is diminishing freedom from death or HVR with increasing time following Kawashima particularly beyond 60 months. Significant risk factors for death or HVR included age at Kawashima where for every 1 month of age increase at time of Kawashima, the Hazard ratio of death or HVR increased by 0.02. The Cox proportional hazard regression demonstrates a reduced early risk ( $p = 0.02$ ) for HVR or death in patients who underwent PA banding versus arterial shunt as the primary procedure. There was an increase in the hazard ratio for death or HVR in patients who had bilateral superior vena cavae.

HVR mortality was relatively high with 2 inter-stage deaths and 4 post HVR. The steepest trajectory of improvement of oxygen saturations is observed in the first 4 months after HVR after which the trajectory plateaus. Time between Kawashima and HVR does not influence the outcome for improvement in saturations. ( $p$  for interaction = 0.221). However, those patients whose time between Kawashima procedure and HVR was more than five years, tended to have lower saturations at time of HVR ( $-6.8\%$ ,  $p = 0.07$ ).

**Conclusion:** These findings advocate elective early HVR post KP in patients with LAI. Those with pulsatile pulmonary artery flow at first surgery could follow a more expectant course but with close monitoring of oxygen saturations to allow for timely intervention.

#### P-111

##### **Timing of the investigations before bidirectional cavopulmonary anastomosis**

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**Objective:** The bidirectional cavopulmonary anastomosis (BDCPA) is a well-established step performed in the single-ventricle pathway. Before accomplishing a BDCPA a cardiac catheterization is routinely performed at our centre. We sought to assess which patients were declined for BDCPA after the initial invasive evaluation and, whether the decision to delay BDCPA resulted in further growth of the pulmonary arteries or amelioration of the hemodynamics.

**Patients and Methods:** 104 consecutive infants with single-ventricle physiology planned for BDCPA were included in this retrospective analysis. 55 patients had a single right, 44 a single left and 5 an undetermined ventricle. A BT shunt had been performed in 41, a Sano shunt in 35 and a central shunt in 7 children; 14 children had a pulmonary artery band and 7 had no previous operations.

**Results:** Median age at the first catheterization was 137 days (range 46–379). Children with Sano shunts were investigated significantly earlier than those with BT shunts (median 125d versus 138d,  $p = 0.01$ ), oxygen saturation did not differ between the shunt types.

In 15 patients (14%) BDCPA was postponed because of small pulmonary arteries ( $N = 7$ ), high pulmonary artery pressure ( $N = 8$ ) or resistance ( $N = 3$ ), high transpulmonary gradient ( $N = 5$ ) or because of an intervention ( $N = 6$ ); 6 of them had a Sano shunt. They were all reevaluated with a second catheterization 19–520 days later (median 131). This investigation did not show additional growth of the central pulmonary arteries (median McGoon 2,0 versus 2,0, Nakata 179 versus 163), but the median transpulmonary gradient was significantly reduced (8 versus 6,  $p = 0.003$ ), while median pulmonary artery pressure (16 mmHg versus 14 mmHg) and resistance ( $3,0\text{WUxm}^2$  versus  $2,0\text{WUxm}^2$ ) tended to be lower. Ventricular function was preserved and atrioventricular-valve regurgitation was not aggravated.

BDCPA was completed by 91 children at a median age of 220 days (range 87–680), with one early postoperative death. Overall interstage mortality was 9.7% (10 children) and 3 patients underwent transplantation.

**Conclusions:** Delaying BDCPA in selected children leads to reductions of pulmonary artery pressure, pulmonary vascular resistance and transpulmonary gradient without signs of impaired ventricular function. However central pulmonary arteries do not seem to grow further.

#### P-112

##### **Noninvasive cardiac output measurement at rest and during exercise in pediatric patients following interventional or surgical ASD-closure**

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**Background:** In the majority of patients secundum ASDs are closed interventionally or surgically prior to the onset of symptoms. Since right ventricular dimensions usually normalize



following ASD closure it has been assumed that cardiac function during exercise will be normal in the long-term follow-up. The inert gas rebreathing (IGR) method allows noninvasive determination of the cardiac index (CI) under exercise conditions. The aim of this study was to determine CI at rest and during exercise in the medium term follow-up in children who underwent a surgical or interventional closure of the defect.

**Patients/Controls:** Seventeen patients (age 8.8–17.3 years) who underwent surgical correction were included together with 17 subjects who received an interventional procedure (age 12.2–17.3 years). The current study was performed 6.5–11.6 years after the procedure in all of them. Twelve healthy children (8.5–18.6 years) served as controls.

**Methods:** The Innocor system is based on the IGR principle. Patients are breathing over a period of 30 seconds a low concentration mixture of an inert and a blood soluble gas from a closed system. The pulmonary blood flow (PBF) absorbs the blood soluble component, the rate of absorption being proportional to the PBF; the higher the PBF the higher the absorption rate will be, while the inert component is expired unaltered. For exercise testing the standard treadmill protocol of the German Society of Pediatric Cardiology was used. CI, stroke volume (SV) and the heart rate (HR) were determined during rest and at two standardized submaximal exercise levels (level 3 and level 6).

**Results:** The CI increased in all subjects under exercise conditions. Neither the SV nor the HR displayed significant differences among the three groups either during resting or exercise conditions. While the HR rose continuously during the exercise test, no increase of the body surface area indexed SV occurred after level 3.

**Discussion:** Noninvasive determination of the CI at rest and during exercise with the IGR is feasible in the pediatric age group. In the medium-term follow-up we found no significant differences between patients who underwent surgical or interventional ASD closure as compared to normal controls.

#### **P-113 Comparison of Electrocardiographic and Echocardiographic/Deformational Imaging Screening in a Cohort of Elite High School Athletes**

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**Background:** The use of electrocardiography (ECG) in athletic screening has been studied widely. In trained athletes, ECG aberrations are common, including increased QRS voltages and repolarization alterations. These findings often prompt evaluation for underlying cardiac pathology. We sought to determine the correlation between atypical ECG findings with results from echocardiography and deformational imaging in a cohort of elite high school athletes.

**Methods:** We prospectively performed a standard 12-lead ECG immediately followed by standard two-dimensional (2D), spectral Doppler, tissue Doppler, and 2D longitudinal strain analyses in 103 high school students (age  $15.9 \pm 1.5$  yrs, 55 males), including 78 highly-trained athletes and 25 healthy non-athlete controls. Longitudinal 2D strain was performed to evaluate 17 regional (apical, mid, and basal) myocardial segments and global longitudinal left ventricular (LV) myocardial strain. ECG-based assessment for left ventricular hypertrophy (LVH) and right ventricular hypertrophy (RVH) was compared to imaging data.

**Results:** Overall, 24/75 (32%) athletes had ECG findings suggesting RVH or LVH (or both), compared to 1/28 (4%)

controls ( $p < 0.01$ ). No patients had significant structural heart disease by echocardiography. LV dimensions and LV mass were significantly greater in athletes ( $p < 0.001$ ). Similarly, ECG deflections in the lateral precordial leads were increased in athletes, including R-wave deflections in lead V6 ( $p < 0.03$ ) and total R+S wave deflection in lead V4 ( $p < 0.02$ ). Traditional R- and S-wave measurements in lead V1 did not correlate in either cohort with increasing LV wall thickness, LV mass, LV end-diastolic diameter, LV tissue Doppler, or global longitudinal 2D strain. Instead, the total R+S wave deflection in lead V4 correlated with LV posterior wall thickness ( $p < 0.0002$ ), LV mass ( $p < 0.008$ ), LV end-diastolic diameter ( $p < 0.0001$ ), and global longitudinal 2D strain ( $p < 0.05$ ).

**Conclusions:** As expected, athletes had a higher incidence of atypical ECG findings compared to healthy controls. However, R- and S-wave deflections in the lateral precordial leads correlated better with echo-measured LV wall thickness and function than lead V1. Determining the overlap between the magnitude of R+S wave deflections in V4 in the elite athlete's heart compared to patients with clinically evident hypertrophic cardiomyopathy may help define a more sensitive and specific ECG screen for this disease.

#### **P-114 Whole-Blood Aggregation Test Stimulated by ADP for Evaluation of Blood Aggregation Activity in Kawasaki Disease Patients with Anti-Platelet Management**

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**Introduction:** It is a very critical issue to evaluate the efficacy of anti-platelet agents by any monitoring system because there is individually any significant difference in responsibility to anti-platelet agents. Recently, the aspirin resistance has been identified as one of the topics. We have already measured the platelet function in patients of Kawasaki disease (KD), and also reported that the analysis with whole blood aggregation have some superiorities over platelet-rich plasma (PRP) aggregation method which is a conventional method. The stimulus was used collagen since we focused on aspirin. However, aspirin is often combined with different types of anti-platelet drugs, thienopyridine, in KD patients with any coronary lesion. This study comprehensively evaluated the effect of all anti-platelet agents administered by whole-blood aggregation using adenosine diphosphate (ADP) which is available for judging the efficacy of aspirin and thienopyridine compound.

**Methods:** The subjects were 48 patients late after KD. Twenty-nine patients had received the anti-platelet therapy mainly with aspirin for coronary artery lesions (CALs), and the remaining not received any medication because they did not have any CALs. Whole-blood aggregation was analyzed using collagen and ADP as the stimulus and was compared with the PRP aggregation measured using collagen as the stimulus with an optical aggregometer. Whole-blood aggregation was evaluated on the basis of the platelet-aggregation threshold index (PATI), which was defined as the putative agonist-concentration giving half-maximal-aggregation. PRP aggregation was categorized into 5 classes; -2, -1, 0, +1, and +2.

**Results:** We found a significant negative correlation between the whole-blood PATI stimulated with collagen or ADP and the PRP aggregation class stimulated with collagen. (collagen: -0.76, ADP: -0.59) In addition, PATI was significantly

decreased in subjects with the combination therapy including any thienopyridine agent when compared with that of all the subjects did not receive any thienopyridine agent. ( $p < 0.01$ )

**Conclusions:** This study revealed that the whole-blood aggregation using ADP as the stimulus contributed to the efficacy of aspirin in KD patients. Furthermore, it may be clinically useful for the comprehensive evaluation of anti-platelet therapy including thienopyridine agents.

#### P-115

##### **Usefulness of NT-pro-BNP in evaluation of the stage of heart failure in infants with ventricular septal defect**

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**Introduction:** Ventricular septal defect (VSD) is one of the most common congenital heart diseases. In some patients hemodynamical disturbances are clinically important and manifestations of heart failure (HF) appear.

**Aim:** The aim of the study was to assess if serum NT-pro-BNP levels correlate with clinical severity of HF in children with VSD.

**Methods:** In 34 children aged 38–338 days (mean  $130 \pm 81$  days), 15 boys and 19 girls VSD was diagnosed by physical examination, ECG, chest X-ray and two-dimensional echocardiography. Pulmonary hypertension was excluded in all patients. HF was diagnosed based on anamnesis and physical examination. All patients were classified by a single observer to the adapted NYHA functional class, the modified Ross score and the New York University Pediatric Heart Failure Index (PHFI). In all of them serum NT-pro-BNP levels were assessed. The results were compared to the control group of 31 healthy children matched for age and sex with the study group. Statistical analysis was obtained by using t-Student test and Pearson's correlation coefficient with the level of significance.

**Results:** The mean values of NT-pro-BNP were significantly higher in the study group than in healthy infants ( $74,9 \pm 73,7$  vs.  $23,7 \pm 21,1$  fmol/l,  $p < 0,0005$ ). In 24 children with VSD signs and/or symptoms of HF were present (failure to thrive in 13, diaphoresis in 9, prolonged feeding time in 5, tachypnoe and dyspnoe in 14, retractions in 22, resting sinus tachycardia in 9, hepatomegaly in 11, decreased peripheral perfusion in 3). In subgroup of children with HF levels of NT-pro-BNP were significantly higher ( $p = 0,001$ ). The values of NYHA grade were: min. 1 – max. 3 (mean  $1,72 \pm 0,74$  points), Ross score: 0–10 ( $4,11 \pm 3,26$ ) and PHFI score: 0–14 ( $5,29 \pm 4,08$ ) and NT-pro-BNP level correlated with the severity of HF:  $r = 0,58$  ( $p = 0,003$ );  $r = 0,51$  ( $p = 0,002$ );  $r = 0,64$  ( $p = 0,00005$ ) respectively. Pharmacological treatment of HF was introduced in 19 infants and 25 were qualified for cardiosternotomy.

**Conclusions:** Serum NT-pro-BNP level correlates to severity of HF in children with VSD and could be helpful in therapeutical approach.

#### P-116

##### **Diastolic function measured by Tissue Doppler Imaging in paediatric patients with End Stage Renal Disease**

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**Introduction:** Cardiovascular disease is the main cause of death in patients with End Stage Renal Disease (ESRD). Early signs of

cardiovascular disease include left ventricular hypertrophy (LVH) and diastolic dysfunction (DD). The aim was to compare the prevalence of LVH and DD in children with ESRD and healthy children using conventional echocardiography and Tissue Doppler Imaging (TDI).

**Methods:** 32 children with ESRD and 79 healthy control subjects, matched for weight or body surface area (BSA), were assessed with conventional echocardiography and TDI. Parameters related to LVH (IVSd%, LVPWd% and LV mass index) and DD (E/a ratio and E/e' ratio) were compared in the ESRD and control groups using linear regression analysis. LVH was defined as LV mass index  $> 103$  for boys and  $> 84$  g/m<sup>2</sup> for girls [1]. DD was defined as E/a ratio  $< 1$  or E/e' ratio  $> 95$  for age.

**Results:** The children with ESRD were significantly older than their healthy controls with the same weight or BSA, mean difference (MD) [95% Confidence Interval (CI)] 2.6 [0.5–4.6] years ( $p = 0.015$ ). After adjustment for age, the ESRD patients had a smaller mean IVS-E (MD [95% CI] 3.6 [2.6–4.6],  $p < 0.001$ ) and MV E/a ratio (MD [95% CI] 0.32 [0.09–0.55],  $p = 0.007$ ) and larger IVSd% (MD [95% CI] 20.9 [14.4– 27.5],  $p < 0.001$ ), LVPWd% (MD [95% CI] 19.7 [12.8–26.7],  $p < 0.001$ ), LV mass index (MD [95% CI] 18.1 [10.9– 25.4],  $p < 0.001$ ) and E/e' ratio (MD [95% CI] 2.5 [1.8– 3.2],  $p < 0.001$ ) than the control subjects. LVH was diagnosed in 3 of the 31 children (10%) with ESRD and none of the controls ( $p = 0.04$ ). DD was diagnosed in none of the children according to the E/a ratio. According to the E/e' ratio 11/26 (42%) of the patients with ESRD and none of the controls were diagnosed with DD ( $p < 0.001$ ).

**Conclusion:** Children with ESRD have significantly more LVH, larger IVSd%, LVPWd% and LV mass index, E/e' ratio and a smaller E/a ratio and IVS-E than healthy, weight or BSA matched, controls. It seems that Tissue Doppler is more sensitive in the detection of DD earlier than conventional E/a ratio in patients with ESRD.

#### P-117

##### **Relationship between Inflammation and Oxidative Stress of Kawasaki Disease in Acute Phase**

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**Background:** In Kawasaki disease, the vicious circle of reactive oxygen species (ROS) and inflammations is formed. The ROS were generated excessively by abnormally appearance of various inflammatory mediators, and these mediators are induced by excessive ROS. We measured reactive oxygen metabolites (ROM) and biological antioxidant potential (BAP) as oxidative stress markers and various cytokines as inflammatory markers to study on the relation between oxidative stress and inflammation. **Methods:** We measured oxidative stress markers (ROM and BAP) and inflammatory markers (hs-CRP, IL-1,2,6, and TNF- $\alpha$ ) in 19 KD patients (0Y10M–5Y10M) in acute phase. 13 patients are favourable and 6 patients are unfavourable to IVIG. Blood samples were obtained before and after IVIG treatment.

**Results:** ROM and all inflammation markers decreased in parallel with the clinical findings in the IVIG favourable group. However, in the IVIG unfavourable group, the value of hs-CRP and IL-6 as sensitive inflammation markers decreased, but ROM and IL-1, 2, did not significantly reduce in values. The value of TNF- $\alpha$  significantly decreased in the IVIG

favourable group, otherwise slightly increased in the IVIG unfavourable group.

**Conclusions:** The difference between two groups in the dynamics of inflammation markers and ROM suggested that oxidative stress may take part in the formation of acute vasculitis through another pathway independent to inflammations, as an indirect inflammation promotion factor. This study also showed that the anti-TNF- $\alpha$  treatment may be useful for IVIG unfavorable patients.

#### P-118

##### Evaluation of left atrial ejection force and ascending aorta elasticity in children after repair of coarctation of the aorta

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**Objectives:** Coarctation of the aorta is a chronic vascular disease characterized by persistency of myocardial and vascular alterations. What remain obscure in these patients are intrinsic mechanisms of hypertension, and its relationship with elastic properties of aorta and left atrium performance. The aim of present study was to evaluate the elasticity of aorta, left atrium function and myocardial performance collectively at midterm follow-up, in normotensive children who have had successful coarctation surgery or balloon dilatation.

**Methods:** In this prospective study, nineteen patients (9 boys, 10 girls; mean age:  $7,15 \pm 0,9$  years) with native coarctation who underwent surgery or balloon angioplasty and 21 age-sex matched healthy children were included. Left atrial ejection force, aortic wall stiffness index, ascending aorta distensibility, M-mode echocardiography, LVMI, diastolic functions and myocardial performance index were assessed in both groups. Left atrial ejection force ( $\text{kdyne/m}^2$ ) is defined as product of mass and acceleration of blood expelled from the left atrium during the accelerative phase of diastole. Aortic stiffness and distensibility were estimated using ascending and descending aorta diameters obtained by M-mode echocardiography and approximation of pulse pressure in the right arm and leg.

Table 1: Relationship between LAEF index and E velocity, E/A ratio as well as right arm systolic blood pressure

Variables	LAEF index	
	p value	r coefficient
E velocity	0.019	0.561
E/A ratio	0.013	-0.586
Systolic blood pressure	0.015	-0.572

**Results:** No difference was found in systolic blood pressure at rest between patients and controls. Left atrial ejection force index were found to be higher in study group than in the control group ( $12.69 \pm 1.76$   $\text{kdyne/m}^2$  versus  $4.57 \pm 1.12$   $\text{kdyne/m}^2$ ,  $p: 0.001$ ). Aortic stiffness index in patient group was significantly increased ( $5.12 \pm 1.24$  versus  $2.57 \pm 0.68$ ,  $p: 0.000$ ). Ascending aorta distensibility was significantly lower in patient group than in the controls ( $42.13 \pm 11.02$  versus  $78.79 \pm 20.49$ ,  $p: 0.000$ ). Correlation was found between LAEF index and right arm

systolic blood pressure, E velocity as well as E/A ratio was found in current study (Table 1).

**Conclusions:** Augmented left atrial ejection index may be indicative of diastolic abnormalities in children who successfully treated in early childhood. We also found that increased aortic stiffness may be used as a marker for late onset hypertension in in the follow-up of coarctation of the aorta.

#### P-119

##### Exercise rehabilitation for children with congenital heart disease

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**Introduction:** The improvement in medical diagnosis and treatment, from pharmacology to surgery, induced an increased survival in children with congenital diseases. In adults with cardiovascular diseases several randomised control trials show beneficial effects of exercise training. Meta analyses have validated the benefits of cardiovascular rehabilitation but none in children with congenital diseases.

**Aims:** Determine the benefits of cardiovascular rehabilitation using Meta analysis methods in children with congenital heart diseases.

**Methods:** Research criteria used were: exercise training, rehabilitation, readaptation, heart disease, heart failure, limited to article in English or French, for subjects' age <25 years and to randomised control trials (RCT) published between January 1966 and December 2010 in database from Pub Med, Embase, Web of Science and CINAHL. All the references were reviewed by 2 independent scientists.

**Results:** 5 RCT studies (n subjects = 173) met the selection criteria. The mean differences reach :  $4,21 [1,69-6,73]$ ,  $p = 0,001$  for maximal oxygen uptake ( $\text{ml.kg.min}^{-3}$ ) ;  $9,4 [3,2-15,6]$ ,  $p = 0,003$  for maximal power (watts) and  $5,9 [-0,4-12,2]$ ,  $p = 0,03$  for heart rate ( $\text{beat.min}^{-2}$ ) at maximal exercise.

**Conclusions:** The results observed in children with congenital heart disease are in agreement and conformity with those observed in adult subjects for the parameters analysed. Exercise training improved significantly maximal oxygen uptake and peak power at maximal exercise. Despite these positive observations, this meta analysis suffer from the limited number of subjects and the lack of data concerning the major acute cardiovascular events which do not permit to extrapolate the entire benefits observe in an adult population. To our best knowledge there is no study evaluating the impact of exercise training in morbid-mortality in a congenital heart disease population from children to adults.

#### P-120

##### Incidence of Pulmonary Artery Complications after Flo Watch Pulmonary Artery Banding

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**Objective:** An assumed advantage of the Flow Watch pulmonary artery (PA) band is that it has a low incidence of PA distortion and requirement for PA reconstruction after its removal. We describe our experience with Flo Watch PA banding with regard to pulmonary complications in a large single-centre population.

**Methods:** A retrospective analysis of all the patients, at our centre, who underwent Flo Watch PA banding to control the pulmonary blood flow for initial single ventricle or bi-ventricle palliation.

**Results:** 56 patients needed Flo Watch PA band between December 2003 and June 2010. 14/56 (25%) had single ventricle morphology and 42/56 (75%) biventricular morphology. Mean age at the time of PA band was 141 (range 7–1486) days and the mean weight 4.7 (range 2.6–15.9) kg. There were 7 deaths in our series, 6 were late deaths and were not associated with PA band. There was 1 early death. 27/56 (48%) had their band removed for next stage surgery and 29/56 (52%) still had the band in place. 18/27 (66%) did not have any PA distortion and did not need any patch enlargement. However, 9/27 (33%) had PA distortion and needed patch enlargement. In 2/27 (7%) the Flo Watch was found to have eroded through the MPA at the time of its removal.

**Conclusion:** Though telemetric Flo Watch PA banding does have undoubted advantages in terms of adjustability of pulmonary flow without reoperation, there is a significant incidence of pulmonary artery distortion requiring patch reconstruction.

#### P-121

##### **Acute Renal Failure Early After Cardiac Surgery With Extracorporeal Circulation: Long Term Outcome?**

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**Background:** Acute renal failure (ARF) in the immediate postoperative period after cardiac surgery requiring extracorporeal circulation (ECC) is common. Long term outcome in pediatric patients is not well known.

**Methods:** We retrospectively reviewed 1407 pediatric cases (age < 16 years) who underwent cardiac surgery requiring ECC, from 1998–2008. ARF was defined as a creatinine value of more than twice the normal value for age occurring within the first 96 hours after surgery.

Current renal outcome was evaluated by means of glomerular filtration rate (GFR) according to Schwartz formula, proteinuria, renal ultrasound with duplex Doppler, blood pressure and cardiologic evaluation.

**Results:** From 1407 patients reviewed, 117 patients (8%) fulfilled inclusion criteria. Pre-existing renal disease was found in 5 patients (4%). Renal replacement therapy (RRT) was required in 5 patients (range 2–45 days). Eighteen patients (15% of study group) deceased due to multiple organ failure or sepsis as the main causes.

The mean follow-up after surgery was 5.6 years (SD 1.6) (FU ongoing). Of subjects requiring dialysis during the immediate postoperative period, none is currently on RRT. Nine patients (31%) have proteinuria. Chronic kidney disease (CKD) was noted in 3 patients, 1 patient stage 1 (GFR 90–100 ml/min/1.73 m<sup>2</sup>), 1 patient stage 2 (GFR 60–89 ml/min/1.73 m<sup>2</sup>) and in 1 patient stage 3 (GFR 30–59 ml/min/1.73 m<sup>2</sup>) CKD with tubular dysfunction. In all three patients small kidneys were noted (SD > -2) with increased corticomedullary reflectivity. Three patients are hypertensive requiring anti-hypertensive treatment, 2 of which have CKD. Renal stenosis was excluded in all patients.

**Conclusion:**

1. Acute renal failure in the immediate post-operative period after pediatric cardiac surgery is not infrequent.
2. Overt Chronic Kidney Disease in early follow-up is limited.
3. Small kidney size and increased corticomedullary reflectivity are specific diagnostic tools for CKD.
4. Very long-term outcome needs to be determined.

#### P-122

##### **The Recognition of Coarctation of the Aorta in Neonates and Small Infants and Its Relation to the Course of the Disease and the Outcomes**

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**Introduction:** Coarctation of the aorta is a common congenital heart disease but may be sometimes missed or underestimated in neonates and young infants. Our institution is the only clinics for pediatric cardiology and cardiac surgery in Latvia. We analyzed all the cases of aortic coarctations operated in our clinics within the first two months of life in the period of time from 2005–2010 to find out the time of diagnosis and its relation to the condition of the patient at the moment of the admission, the course of the disease and the outcomes. The birth rates in our country within years 2005–2010 were 22 422 +/- 947 newborn infants/per year.

**Methods:** Retrospective analysis of the case histories of all the neonates and infants in the age group up to two months undergone surgical correction of the coarctation in our institution between January 1, 2005 and December 31, 2010.

**Results:** 45 neonates and infants presented with aortic coarctation. 27% of the cases were detected antenatally. 64% of the patients were sent by maternity hospital, but 36% after the discharge home (by general practitioner or hospital). In 73% (n = 33) cases congenital heart disease, but in 27% of the cases other diagnoses suspected (sepsis, pneumonia, feeding disturbances etc.). PgE1 was used in 100% antenatally suspected, but 48% postnatally detected cases, the need of inotropes in 8% prenatally, 21% postnatally detected cases. From antenatally detected cases 50%(n = 6) were combined with complex cardiac lesions and the half of them with hypoplastic aortic arches, 25% (n = 3) with VSD. There were 6 lethal cases (3 -heart insufficiency, 2 -septicemia, 1 -renal insufficiency).

**Conclusions:** Most antenatally detected coarctations are combined with other cardiac lesions and/or hypoplastic aortic arch therefore carries higher mortality rates. Postnatally after the discharge from maternity hospital detected cases correlate with older age, more frequent need of inotropes and ventilation, but with lower overall mortality rates. 27% of unrecognized cases in pre hospital stage are indicative of the need for further education for general practitioners and pediatricians working with neonates and infants, because delayed diagnosis may carry worsened surgical outcomes and increased length hospital stay.

#### P-123

##### **Are preoperative left heart dimensions in infants with coarctation repair predictive of long-term outcome?**

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**Introduction:** Infants with coarctation often present with a dilated, pressure overloaded right ventricle and a small left ventricle. Preoperative determination of the adequacy of the left ventricle to support the systemic circulation can be difficult. Left ventricular dimensions are expected to increase rapidly after coarctectomy, mainly as a result of normalization of biventricular



loading conditions. Longterm data on growth of left ventricular structures in this patient group are scarce. The aim of this study was to evaluate long-term outcome in relation to echocardiographic parameters at presentation.

**Methods:** All infants under 3 months of age, who underwent coarctectomy between December 1987 and December 2007 were studied retrospectively. Patients with mild aortic stenosis (gradient < 20 mm Hg) or with ventricular septal defect were included. Clinical charts and first and latest echocardiograms were reviewed. Preoperative diameters of mitral and aortic valve annulus as well as dimensions and length of LV and RV were obtained and compared with the same measurements at last follow-up. Values were expressed as z-scores.

**Results:** A total of 194 infants were included. 32 (16%) underwent concomitant VSD closure, 9 (5%) pulmonary artery banding. Follow-up was 99% complete with a mean of 11,4 years. Early mortality was 2,5%, late mortality 3,6%. During follow-up, recoarctation occurred in 30 patients (16%), 12 patients (6%) required intervention for (sub)aortic stenosis, 7 patients (3,7%) mitral valve surgery, 2 patients developed pulmonary hypertension, because of restrictive LV-physiology. Before coarctectomy, a mitral valve z-score < -2 was present in 10% of the patients, an aortic valve z-score < -2 in 23% and < -3 in 9%. In 75 patients, the ratio RV/LV internal length was < 1. At latest follow-up, only 1 patient remained with a mitral valve z-score < -2 and 10 with an aortic z-score < -2. LV length was  $\geq$ RV length in all. Initial aortic annulus z-scores correlated with the need for aortic/subaortic surgery during follow-up (p = 0,017 t-test).

**Conclusion:** Small left heart structures increase significantly in size after coarctation repair in infancy. A smaller initial aortic valve annulus z-score is predictive for (sub)aortic surgery during long-term follow-up.

#### P-124

##### **Propranolol in the treatment of infantile haemangioma**

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**Background:** Infantile hemangioma (IH) is one of the most common tumoral lesions in childhood. There are several medical or surgical therapeutic options, all with sub-optimal results. Recently, it was described the successful use of propranolol for involution of IH. We report the results of a multicenter experience with this therapeutic option.

**Objective:** Prospective evaluation of efficacy and safety of propranolol for the treatment of IH.

**Methods:** Prospective evaluation of clinical data of all patients with IH referred to a Paediatric Cardiology center during the year 2010 to start treatment with Propranolol. Efficacy was evaluated as a reduction of the lesions. Safety was evaluated by collecting data regarding significant secondary side effects (hypoglycemia, hypotension, bradycardia, drowsiness). A baseline cardiac evaluation included ECG and echocardiogram. Propranolol was started as an outpatient and titrated to a target dose of 2–3 mg/kg/day according to clinical response.

**Results:** During the year 2010, 12 patients (6 female) were referred for propranolol treatment of IH, at a mean age of 4 months (min 1; max 9). Baseline blood pressure, fasting glycemia, heart rate and ECG were normal in all patients. Four had benign associated heart disease: three atrial septal defects and one patent ductus arteriosus. The mean initial dose was 1,1 mg/kg/day, titrated to a mean maximum dose of 2,7 mg/kg/day.

With a mean follow up of 8 months (min 4, max 13). All patients showed a significant reduction of the IH dimensions. There were no side effects.

**Conclusions:** Our preliminary experience confirmed Propranolol as a successful and safe treatment for IH. We believe that at this point, a cardiac evaluation is warranted due to potential associated cardiac defects and secondary effects. Larger studies are necessary to confirm this approach before widespread use without involvement of Pediatric Cardiologists.

#### P-125

##### **Predicting surgical outcome in children with acyanotic congenital heart diseases and severe pulmonary hypertension**

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**Background:** Elevated pulmonary vascular resistance is a contraindication for surgical repair for left to right shunts.

**Objective:** To investigate if perioperative risk stratification be predicted clinically only.

**Methods:** Clinical, surgical, and post-surgical records of patients with complete atrioventricular septal defect (CAVSD) and ventricular septal defect (VSD) who underwent catheterization from 2001 to 2009 were reviewed. Cohort divided into; group A (low risk) having PVR  $\leq$  6 Woods units and group B (high risk) with PVR > 6 Woods units on room air. Standard operability criterion was exercised to judge the operability.

**Results:** Eighty four patients; (VSD = 47, CAVSD = 37) underwent diagnostic catheterization. Mean age/weight =  $3.5 \pm 2.9$  yrs and  $11 \pm 5.7$  kg in VSD group and  $2.9 \pm 3.8$  yrs and  $10.5 \pm 9.3$  kg in CAVSD. Amongst VSD patients, Group A = 23 and B = 24. In CAVSD patients, Group A = 21 and B = 16. All VSD and 33 CAVSD were deemed operable. Mean pulmonary artery pressure and PVR was significantly higher in group B in both VSD. ( $62.5 \pm 13$  mmHg Vs.  $47 \pm 13$  mmHg, p = 0.005 and  $9.3 \pm 3.3$  WU vs  $3.2 \pm 1.3$  WU, p = 0.005) and CAVSD. ( $62 \pm 9$  mmHg vs.  $42 \pm 13$  mmHg, p = <0.001 and  $10.3 \pm 4$  WU vs.  $3.2 \pm 1.6$  WU, p = 0.001). All but three with VSD had SaO<sub>2</sub>  $\geq$ 94%. In CAVSD patients, all meeting the PVR based operability criteria had SaO<sub>2</sub>  $\geq$ 83%. Four inoperable cases had SaO<sub>2</sub> < 80%. Six patients did not undergo surgical repair, and other 3 had missing records. There was no significant difference in tested variables between the two groups of both VSD and AVSD. Two (4%) VSD and 1 (4%) CAVSD patients died within 30 days of surgery. One high risk VSD patient died two years after. One patient in each group was on anti-pulmonary hypertensive medicines. Rest were doing well over a mean follow up of 4.8 years.

**Conclusion:** SaO<sub>2</sub> of 94% in children with VSD and of 83% with CAVSD correctly identifies operable patients. This criterion may be used as an alternate to catheterization to identify operability amongst patients with left to right shunts. Immediate post-operative and short term outcome of operable patients having higher PVR is similar to those with PVR < 6 WU.

#### P-126

##### **The right ventricular growth after decompression in patients with critical right ventricular outflow tract obstruction**

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**Objectives:** To evaluate the right ventricular growth of patients in whom the initial management was right ventricle decompression. Patients with RV dependent coronary circulation were excluded from this study.

**Methods:** Between May 1994 to May 2010, 48 patients (27 M, 21 F) with critical pulmonary stenosis (CPS) or pulmonary atresia with intact ventricular septum (PAIVS), (19 PAIVS, 29 CPS) underwent RV decompression. The mean age at first intervention was  $4 \pm 4,3$  days (range 1–19 days). Mean weight was  $3,05 \pm 0,5$  kg (range 1,9–4,6 kg). Right ventricular development was assessed using tricuspid valve dimensions (TV) and RV length. TV was measured retrospectively on the cross-sectional echocardiograms performed before the procedure and during follow-up. Z score were used to standardize tricuspid valve dimensions with body size. The RV length was assessed from the tricuspid valve annulus to the RV apex at ventricular end diastole. At the latest review, we have evaluated 32 patients (17 CPS, 15 PAIVS). TV valve diameter at birth in these 32 patients ranged from 8 to 18 mm ( $11,5 \pm 2,68$ ), Z score of the TV valve ranged from  $-2,3$  to  $2,87$  ( $-0,08 \pm 1,3$ ).

**Results:** During the follow-up, 29 patients out of 32 have a biventricular circulation, 2 patients have undergone 1 and  $\frac{1}{2}$  ventricle repair, and 1 patient has undergone Fontan operation. Twelve of 32 patients (Group 1) required additional transcatheter or surgical procedures to augment the pulmonary blood flow. For group 1, TV diameter at birth ranged from 9 to 18 mm ( $12,3 \pm 3,3$ ). Z value ranged from  $-1,9$  to  $2,87$  ( $0,3 \pm 1,5$ ). During the follow-up, TV diameter ranged from 19 to 31 ( $23,5 \pm 4$ ). Z score ranged from  $-6,1$  to  $-0,48$  ( $-3,4 \pm 1,6$ ). 29 of 32 patients (group 2) did not require additional procedures. For group 2, TV diameter at birth ranged from 8 to 14 mm ( $11,1 \pm 2,1$ ). Z score ranged from  $-2,3$  to  $1$  ( $-0,3 \pm 1$ ). During the follow-up TV diameter ranged from 20 to 37 mm ( $27,3 \pm 4,2$ ). Z score ranged from  $-3,5$  to  $2,02$  ( $-1,7 \pm 1,5$ ). RV length for group 1 ranged from 21 to 52 mm ( $37,6 \pm 10,8$ ) and for group 2 it ranged from 26 to 52 mm ( $39,6 \pm 8,5$ ).

**Conclusion:** Our data demonstrate that even in cases in which biventricular circulation can be achieved the growth of the right ventricle is not normal. It is important to investigate other factors such as genetic determinants that could influence right ventricular development.

#### P-127

##### **Serum Uric Acid Levels in Normotensive Children With Family History of Essential Hypertension**

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Many factors, including family history, genetics, insulin resistance and high body mass index may play a role in the development of essential hypertension. The family history of hypertension is an important risk factor for essential hypertension seen in children. It has been said that hypertensive patients with nondipper blood pressure status have higher end organ damage. Increased left ventricular mass is an important marker for hypertensive cardiac injury. It has been reported that elevated serum uric acid concentrations contribute development of essential hypertension at early stages. The aim of this study was to evaluate diurnal blood pressure variation (dipper and nondipper status) of normotensive children with family history of essential hypertension and relationship between left ventricular mass and serum uric acid level. The children aged between 8–22 years were enrolled. Forty (19 girls, 21 boys) of them were the

normotensive offspring of hypertensive parents and twenty (10 girls, 10 boys) of them were the normotensive offspring of normotensive parents. Medical history has been obtained, physical examination, 24h ambulatory blood pressure monitoring, echocardiographic examination and calorimetric enzymic measurement of serum uric acid have been performed. Children with family history of hypertension divided into two subgroups according to blood pressure variation as dipper and nondipper group. The nondipper group showed higher left ventricular mass index and serum uric acid levels compared with dippers and controls ( $p < 0.001$ ). Left ventricular mass index results of children with family history of hypertension were corrected by age and then Ridge regression analysis was performed. It is observed that left ventricular mass index change with serum uric acid levels ( $p < 0.001$ ), daytime systolic blood pressure levels ( $p < 0.001$ ), night systolic ( $p:0.006$ ) and diastolic blood pressure levels ( $p:0.029$ ). We conclude that cardiac injury begin much more earlier and its related to elevated uric acid concentrations in the nondipper normotensive children with family history of hypertension.

#### P-128

##### **Comparison of perioperative C-reactive protein between neonates after the Norwood procedure or arterial switch operation**

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**Objectives:** C-reactive protein (CRP) is a widely used indicator of systemic inflammatory response in patients with cardiovascular diseases, including heart failure and after cardiopulmonary bypass (CPB). Neonates undergoing the Norwood procedure (NP) have a poorer systemic hemodynamic and oxygen transport status than other CPB surgeries. We compared CRP levels between the two groups of neonates after NP or arterial switch operation (ASO). **Methods:** Charts of 170 neonates from 2003–2009 were reviewed ( $n = 89$  in NP group,  $n = 81$  in ASO group). CRP was measured in 66 neonates in NP group and 47 in ASO group twice weekly and recorded prior to and within 20 days after CPB. White blood cells, doses of inotropes and steroid, cultures of blood and body fluids were recorded. Demographic data included the durations of CPB, aortic cross clamp and circulatory arrest, ICU and hospital stay.

**Results:** NP group had a shorter CPB. ( $109 \pm 39$  min) and aortic cross clamp ( $44 \pm 19$  min) than ASO group ( $143 \pm 76$  min and  $75 \pm 26$  min) ( $p = 0.002$  and  $p < 0.0001$  respectively), but a longer circulatory arrest ( $22 \pm 11$  min vs.  $8 \pm 11$  min,  $p < 0.0001$ ). Prior to CPB, CRP was higher in NP group than ASO group ( $21.6 \pm 24.0$  mg/L vs.  $13.1 \pm 25.6$  mg/L,  $p = 0.01$ ). CRP increased to  $80.2 \pm 47.7$  mg/L in day 1–2 after NP and  $73.2 \pm 35.8$  mg/L after ASO ( $p < 0.0001$  for both groups), then gradually decreased to  $40.9 \pm 35.7$  mg/L in NP group and to  $20.0 \pm 18.6$  mg/L in ASO group by day 20 ( $p < 0.0001$  for both groups). Throughout the postoperative period, CRP was significantly higher in NP group than ASO group ( $p = 0.036$ ). When the two group data were analyzed together, CRP was significantly and positively correlated with neutrophil count ( $p = 0.0008$ ) and negatively correlated with lymphocyte count ( $p = 0.008$ ), not with other variables.

**Conclusions:** Neonates undergoing NP have a greater systemic inflammation before and after CPB than those undergoing ASO, despite shorter CPB and ACC. This may indicate an important

role of systemic hemodynamics and oxygen transport status in systemic inflammatory response in addition to CPB in neonates with congenital heart defects.

#### P-129

##### **Is Hypoplastic Left Heart Syndrome a Disease of Aortic Valve? A Genetic Perspective**

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**Objective:** Hypoplastic left heart syndrome (HLHS) is one of the most severe congenital heart malformations, characterized by a large spectrum of underdevelopment of the structures in the left heart-aorta complex. One of the challenges in the management of these patients is the decision to proceed with a 1- versus 2-ventricle repair. Several observations suggest that in some patients the ventricle is still capable of growth when the primary defect is in valve development, while in others HLHS may be due to a primary defect in left ventricular development and growth. The aim of this study was the identification of the specific genetic causes of HLHS that may lead to a stratification scheme facilitating the selection of surgical strategies.

**Methods:** We have analyzed 53 well-characterized patients, using an integrated genomic approach combining DNA sequencing of five candidate genes and a genome-wide survey by high-resolution array CGH.

**Results:** We have identified in 30 patients two de novo mutations in NOTCH1, 8 rare inherited gene variants in NOTCH1, FOXC2 and FOXL1 and 33 mostly inherited copy numbers variants. Some of these variants coexist in the same patient.

**Conclusions:** Our findings predict that HLHS is characterized by a complex and heterogeneous pattern of inheritance with rare de novo highly penetrant mutation or multiple interacting low penetrant alterations contributing to the etiology of the disorder. Moreover, in-silico analysis of the identified anomalies shows a functional association between seven genes involved in cardiac valve development indicating that HLHS is at least in part a "valve" disease.

#### P-130

##### **Etiological and Clinical Aspects of Dilated Cardiomyopathy in Children in North-Eastern Romania**

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**Introduction:** Dilated cardiomyopathy is the most common type of heart muscle disease in children. The reported incidence rate is 0.57 cases per 100,000 children, but studies are not available in most countries. Causes of this condition include: myocarditis, neuromuscular disorders, nutritional deficiencies, collagen vascular diseases, hematological diseases, coronary artery diseases, certain types of medications, endocrine and metabolic disorders and certain malformation syndromes. The goal of this study is to establish the causes that led to the occurrence of dilated cardiomyopathy in 21 children, aged 3 months-18 years, hospitalized in Ist Pediatrics Clinic of "Sfanta Maria" Emergency Children's Hospital from Iasi, Romania, to describe the clinical presentation and laboratory workup that allowed us to establish this diagnosis. The children were hospitalized from January 2007 to January 2011. Medium age at the time of diagnosis was 10,95 years.

**Methods:** We have performed a prospective analysis including physical examination, EKG monitoring and echocardiographic examination on all patients. Depending on the associated pathology, other tests were also necessary.

**Results:** We have encountered 1 case of progressive muscular dystrophy (Duchenne), 2 cases of Werdnig-Hoffman disease, 2 cases of Hodgkin malignant lymphoma, 1 case of non-Hodgkin malignant lymphoma, 1 case of malignant germinal tumor, 4 cases of chronic renal failure (undergoing hemodialysis), 1 case of etnobotanic substance intoxication, 2 cases of sepsis, 3 cases of acute myocarditis and 4 cases of lower respiratory tract infection. 16 patients (76%) had signs and symptoms of heart failure (various NYHA stages). 7 patients (33%) died due to their heart condition.

**Conclusion:** The etiology of dilated cardiomyopathy in children is diverse, and its outcome, symptoms and severity depend on the cause and associated pathologies. Symptoms of heart failure represent a major prognostic factor for the need of heart transplantation.

#### P-131

##### **Alström Syndrome—a rare disease presenting with dilative cardiomyopathy and blindness. Two case reports**

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**Introduction:** Alström Syndrome (AS) is a rare, autosomal recessive single gene disorder with varying clinical features including blindness/ early nystagmus, dilative cardiomyopathy, hearing loss, obesity, Type 2 diabetes mellitus, hepatic dysfunction and renal failure. Diagnosis can be confirmed by analysis of mutations in the ALMS1 gene on chromosome 2p13. Only symptomatic therapy is available. We report on two cases: A 12 year old, blind boy that presented with acute symptoms of cardiac decompensation that led to cardiopulmonary resuscitation and death. Further about a two year old girl with neonatal cardiomyopathy, nystagmus and ventricular septal defect now presenting with severe photophobia during admission for VSD- Repair.

**Methods:** Case report, review of literature.

**Results:** Because of the combination of cardiomyopathy and blindness we suspected Alström syndrome in both cases. Genetic analysis in both patients revealed mutations in the ALMS1 Gene.

**Conclusions:** Neonatal cardiomyopathy in combination with vision impairment and nystagmus are leading symptoms for Alström syndrome. The diagnosis can be confirmed by genetic analysis of the ALMS1 gene.

#### P-132

##### **The evaluation of the long-term effect of intrauterine malnutrition on the cardiac functions**

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Intrauterine growth retardation is predisposed cardiac dysfunction. The aim of this study was to investigate the impact of intrauterine malnutrition on the ventricular functions in the small for gestational age (SGA) babies with the long-term postnatal age.

The cardiac functions of twenty term asymmetric SGA infants and 20 term appropriate for gestational age infants were prospectively evaluated by conventional and tissue Doppler

echocardiography (TDI) in their postnatal fifth day, at the end of first month and the third month.

There were no differences between the groups in gender, blood pressure and heart rate ( $p > 0.05$ ). Although, Left ventricle end-diastolic diameter, posterior wall thickness, interventricular septum thickness and left ventricle mass significantly increased with increasing age in the both groups, all these parameters were significantly lower in SGA infants ( $p < 0.05$ ). No differences were determined systolic and diastolic functions by the conventional echocardiography ( $p > 0.05$ ). Peak systolic velocity at fifth day and early diastolic velocity in all evaluations determined by TDI for left ventricle (MSa, MEa) were significantly lower in SGA group ( $p < 0.05$ ). E/MEa ratio was significantly higher in SGA infants at all evaluations ( $p < 0.001$ ). Peak systolic velocity, early diastolic velocity, and Ea/Aa ratio for the interventricular septum (IVSSa, IVSEa, IVSEa/IVSAA) were found to be lower in SGA group at all evaluations ( $p < 0.05$ ). Early diastolic velocity, and Ea/Aa ratio for the right ventricle (TEa, TEa/TAA) were found to be lower in SGA babies at all evaluations ( $p < 0.05$ ). E/TEa ratio was found to be higher in SGA at all evaluations ( $p < 0.001$ ). Sa, Ea, Ea/Aa ratio for the right ventricle, left ventricle and interventricular septum were increased with increasing age ( $p < 0.001$ ). A positive correlation was detected between the ponderal index and Ea for the right ventricle, left ventricle and interventricular septum ( $p < 0.05$ ).

Intrauterine growth retardation is associated with the increased risk of cardiovascular disease in adult life; it is unclear whether the relationship is present at infant's period. These data suggest that indexes of diastolic function, including tissue Doppler measures, were significantly impaired in SGA babies at the third month after birth.

#### P-133

##### **Palliative Potts anastomosis for primary pulmonary hypertension in children: mid-term results**

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Despite permanent progresses of medical treatments, primary pulmonary hypertension in children (PPHTC) remains a not curable disease with a severe prognosis. Moreover, permanent intravenous treatment is particularly unacceptable for the quality of survival at this age.

**Background:** In May 2004, it was decided to try a surgical palliative treatment: anastomosis from the descending aorta to the left pulmonary artery without cardiopulmonary bypass. The reason of this attempt was the relatively good prognosis of Eisenmenger syndrome with large patent ductus arteriosus.

**Objectives:** To know the risks of that surgery and the mid term results of the first cases.

**Methods:** From 06/05/2004 to 23/03/2007, six children underwent Potts anastomosis for PPHTC. Age were from 2.4 to 11 years, weight from 14 to 23 Kg. All were NYHA IV. All received Bosentan, all but two intravenous permanent prostacyclin and one Revatio.

**Immediate Results:** No death occurred during operation.

One child died at D12 with staphylococcus infection and major cyanosis: it was the child with Bosentan monotherapy.

**Mid term Results:** No death occurred during a mean follow-up of 4 years and 2 months. For the 5 surviving patients, functional status increased from NYHA IV to NYHA I. (3 pts) and NYHA II. (2 pts). Intravenous prostacyclin therapy was stopped for 3/4 pts who received it before. Potts anastomosis remained large with right to left shunt and same pressure in pulmonary arteries and aorta. Oxygen saturation in inferior limbs is stable, from 88 to 72%. All have mild to moderate polycythaemia.

**Conclusion:** Surgical Potts anastomosis is a palliative solution for PPHTC with an acceptable perioperative risk and a good midterm result.

#### P-134

##### **Arterial Hypertension in Children in in North-Eastern Romania**

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**Introduction:** Hypertension, a major cardiovascular risk factor, is still a major public health problem worldwide. Unlike in adults, the incidence of hypertension in children is not known, due to the lack of studies for this specific population. Still, hypertension is more and more frequently diagnosed in children. In this population, hypertension is rarely essential, therefore if the cause can be determined, it can often be cured. This is important because untreated hypertension correlates with heart failure, myocardial infarction or stroke in the adult-to-be. The goal of this study was to describe the profile of the hypertensive pediatric population in north-eastern Romania.

**Methods:** We have performed a retrospective analysis of pediatric patients hospitalized in the Pediatric Cardiology Ward of "Sfanta Maria" Emergency Children's Hospital in Iasi, Romania, diagnosed between July 2005 and June 2010 with arterial hypertension. The study protocol included age, sex, height, weight, body mass index, history of the disease, clinical examination, nutritional evaluation, urinalysis, biochemical parameters, lipid profile, renal ultrasound, echocardiography, endocrinological examination, angioCT, blood pressure monitoring.

Blood pressure values were interpreted according to the latest child hypertension classification "The Fourth Report on the Diagnose, Evaluation and Treatment of High Blood Pressure in Children and Adolescents"- 2007. Hypertension was defined as SBP/DBP  $\geq$  95th percentile (after 3 measurements).

**Results:** 83 patients (51 boys and 32 girls) were identified to have arterial hypertension; children were aged  $13,68 \pm 4,2$  years. 41 children (49,3%) were obese. 3 patients (2 girls and 1 boy) were diagnosed with coarctation of the aorta. One patient was diagnosed with adrenogenital syndrome and another one with neurofibromatosis. Urinalysis and biochemical parameters were normal in all cases. 17 children (20%) had elevated total cholesterol levels and 9 patients (10,8%) had severe hypercholesterolemia ( $>200$  mg/dl). 3 patients (3,6%) had renovascular abnormalities. The diagnosis of coarctation of the aorta was established by angioCT in 3 cases.

**Conclusions:** The most important risk factors associated with arterial hypertension were obesity, hypercholesterolemia and heart malformations. For children and adolescents with hypertension, initial evaluation should consist of ambulatory blood pressure monitoring, cholesterol level measurement, lipidic profile, renal ultrasound, endocrinological examination and echocardiography.

#### P-135

##### **Sildenafil: Experience in Children with or without Pulmonary Hypertension and Congenital Cardiac Defects**

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**Background:** Data regarding dosage, safety, and clinical and hemodynamic responses to Sildenafil for management of patients with congenital heart disease (CHD) with or without pulmonary artery hypertension (PAH) has only been reported sporadically.



**Aim:** Describe the cohort of children in whom Sildenafil was used and determine the clinical and corresponding hemodynamic response.

**Methods:** We retrospectively studied 85 patients (108 records) who were treated between January 2008 and December 2010 (mean age 4.2 years; 42 females; mean weight 13.5 kg). Patients were classified into 4 groups; 1: Idiopathic PAH (N = 8); 2: Biventricular circulation with PAH, including Cardiomyopathies and CHD (N = 30); 3: Right ventricular dysfunction (Fallot type and Heart transplantation, N = 9); 4: Univentricular circulation (N = 59). We recorded dosage, duration, side effects, and setting of treatment: Outpatient (N = 4); Hospitalized (N = 47); Post-heart surgery (N = 56). The following objectives were analysed: 1) Respiratory improvement (withdrawal of mechanical ventilation, supplementary oxygen or nitric-oxide); 2) Echocardiographic improvement (PAH estimation and ventricular function); 3) Clinical situation (oxygen saturation and functional status); and others, such as decrease of debit drains on starting, at 6 months or on terminating treatment. Hemodynamic response in those undergoing catheterization pre (N = 85) and post (N = 65) treatment was analysed.

**Results:** The mean initial dosage was 1.68 mg/kg/d (Range 0.4–4) and mean duration of treatment was 12.6 months. Escalating to the maximum dosage occurred within 48 hours in 65.4% (mean maximum dosage 2.36). Side effects were reported in 17 patients, requiring withdrawal in 2 (1.9%) both postop. An improvement was observed in at least one objective on starting in 66 records (70%), especially in respiratory evolution 42.4%. Improvement in pulmonary vascular resistance and mean pulmonary pressures was observed in Groups 1, 2 and 4 ( $P < 0.05$ ). Improvement in flow ratio (Qp/Qs) was observed in Group 3 ( $P < 0.05$ ). The children with echocardiographic ventricular dysfunction pretreatment (N = 28) had a worse initial response ( $P = 0.006$ ), except for patients in Group 4 (acute right ventricular dysfunction postransplant, N = 4) which improved. **Conclusion:** Oral administration of Sildenafil in children with or without PAH and CHD is safe and has a favourable clinical and hemodynamic response with the use of standard dosage, except in those patients with worse ventricular function.

### P-136

#### Patient Registry of the Competence Network for Congenital Heart Defects in Germany

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**Introduction:** Many CHD patients remain chronically ill throughout their lives and require lifelong care. Since patient numbers are small and due to the high variability of disease patterns, there is a lack of sufficient data for the development of adequate treatment and care. Therefore in 2003 the German cardiac associations have initiated a patient registry that collects data and (since 2008) biomaterial samples from patients of all ages and from all over Germany on a long-term basis in a central database & biobank facility.

**Methods:** Patients (or parents of underage patients) enrol on their own account via a registration form that is distributed by most German heart centres and parents' associations. On registration and entry of personal data (IDAT-database), a unique patient identifier (PID) is generated. The PID applies for all databases within the Competence Network (including a clinical study- and image database) and ensures a correct assignment of data sets. Health-related and medical data are based on patient questionnaires and physician's medical reports and are stored in a separate database (MDAT-database) to ensure data privacy.

Diagnoses and procedures are classified using the EPC-Code/Short List. EDTA-blood samples are collected via (i) direct contact to registry patients and their families, and (ii) cooperation with several involved clinical centres in Germany. DNA isolated from EDTA-blood is stored in a central facility.

**Results:** Presently (January 2011) the registry comprises a total of 40,013 patients. The large majority (84%) of participants are underage, sex is evenly distributed. Simple heart lesions (VSD, ASD, PDA) represent 51%, moderate (TOF, ISTA, AoV, TGA, AVSD, PaV, DORV, PAPVC, TAPVC, HRST) 42%, and severe/complex (DIV, CMP, PA, CCT, TAC, HLHS, Marfan) 7%. Based on registry data and directed recruitment of participants numerous studies have been initiated regarding quality of life, social situation, patient communication, genetic causes, as well as clinical studies. The DNA collection presently comprises 712 samples covering a wide range of CHD phenotypes. The sample collection will be extended to include also cardiac tissue, a pilot phase has started in autumn 2010.

**Conclusions:** The patient registry and biobank facilitates collaborative and translational research on congenital heart defects.

### P-137

#### Sildenafil as preparation for the Fontan Procedure

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**Introduction:** Patients with Glenn procedure must have optimum pulmonary pressures and pulmonary vascular resistance to face Fontan physiology. Sildenafil has been used in the treatment of primary and newborn pulmonary hypertension, but its role in the single ventricle is not fully understood. We describe our experience with this drug in treatment for optimization of hemodynamic values in patients before bicavopulmonary anastomosis.

**Methods:** 23 patients, mean weight of 17,074 g, and with mean age of 5.4 years. All of them with anatomical substrate of single ventricle with aortic hypoplasia and Norwood and Glenn procedures performed.

First catheterization was done for anatomic and hemodynamic study. Pulmonary vascular resistance index (PVRI), mean pulmonary pressure (PAP), wedge pressure (WP), and right ventricle end diastolic pressure (RVEDP) were measured. In 15 patients aortopulmonary and veno-venous collaterals were embolized with coils and vascular plug devices (figure).

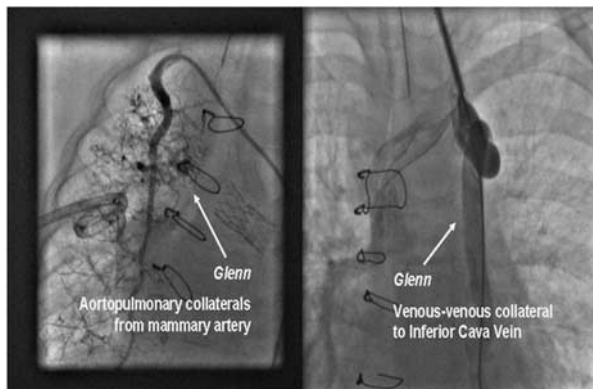
The indication for Sildenafil administration was the existence of mean PAP > 12 mmHg, pulmonary vascular resistance index > 1.5 units Wood/m<sup>2</sup>, and/or the existence of veno-venous collaterals that could work like a discharge circuit of the upper venous territory (fig.). The treatment was maintained a minimum of 6 months (median 11.4) at a dose of 1–4 mgrs./kilo/day (mean, 1.87 mgms/kilo/day) until Fontan procedure.

**Results:** Catheterization prior to surgery measuring the same hemodynamic parameters was performed, with the following comparative results:

	1° Catheterization	2 <sup>nd</sup> Catheterization	P
PVRI (U.Wood/m <sup>2</sup> )	1.06	1.03	0.07
Qp/Qs	0.59	0.91	< 0.02*
WP (mmHg)	10.4	8.7	0.09
RVEDP (mmHg)	10.9	10.2	0.2
PAP mmHg	12.7	11	0.08

After treatment with Sildenafil, an improvement in all hemodynamic parameters was observed, although statistical significance was only obtained in Qp/Qs increase ( $p < 0.02$ ).

Fontan was completed in 6 of these patients without mortality. Empirically the treatment was maintained during the next 6 months after surgery.



**Conclusions:** Pretreatment with sildenafil before Fontan surgery, can improve hemodynamic parameters of pulmonary circulation, improving mortality and morbidity in the postoperative period, specially in risk patients.

#### P-138

##### The PAN study: Association of Gestational and Peripartal Characteristics with Different Cardiovascular Malformations

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**Objectives:** From July 2006 until June 2009 about 20.000 live births with CHD have been registered in the nationwide PAN-Study ("Praevalenz angeborener Herzfehler bei Neugeborenen") in Germany. Recently, we have documented the prevalence of the different congenital cardiovascular malformations. In this report, the focus is on pre- and postnatal diagnostic assessment of the CHD as well as on the association with pregnancy related data, such as maternal age, gestational age and birth weight of the patients.

**Results:** According to the parents' reports, a prenatal echocardiography has been performed in 31.5% of all pregnancies. Frequency of prenatal detection of the CHD strongly correlated with the severity of the cardiac defect. Among the PDA dependent lesions, which accounted for 10% of all CHD cases, the prenatal diagnosis was made in 96% of the univentricular hearts, 80% of PA/VSD, 92% of IAA and in 55% of CoA. Postnatal diagnosis was ascertained in 95% of the PDA dependent CHD patients.

Compared to all live births in Germany during the study period, multiple births, prematurity and low birth weight were doubled in infants with CHD. Prematurity and low birth weight were associated with multiple pregnancy in 69% and 47%, resp, and to a less degree with genetic and extracardiac anomalies. As expected, the association of multiple births was highest with PDA and ASD, but there was a correlation with Ebstein anomaly and valvular PS as well.

About 15% of all patients with CHD were small for gestational age babies. The SGA-risk was highest for PA/VSD, TAC, ToF and DORV.

The maternal age of all CHD cases was similar to that of live births in the general population; it was slightly elevated in CHD patients from multiple pregnancies and significantly elevated in CHD and additional genetic anomalies. Of all cardiovascular defects, only the Ebstein anomaly was found to be associated with a maternal age  $>33$  years.

**Summary:** Severe, PDA-dependent cardiac lesions were effectively detected by pre- as well postnatal echocardiography. Multiple births, prematurity and low birth weight were doubled in patients with CHD.

(Study support by the German Federal Ministry of Education and Research, FKZ01GI0601).

#### P-139

##### Echocardiographic and electrocardiographic parameters as predictors of a worse outcome in children with hypertrophic cardiomyopathy

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**Background:** The clinical presentation and natural history in children with HCM is heterogeneous, ranging from asymptomatic forms to malignant expressions that may result in sudden cardiac (SCD) or heart failure-related death. The aim of study was to assess the prognostic value of echocardiographic parameters in addition to clinical and electrocardiographic characteristics in children with HCM.

**Methods:** Retrospective analysis of 88 pts, mean age 10,6 yrs with HCM diagnosed from 1991 to 2010. Mean follow-up was 6,9 yrs. All 88 pts were divided into two groups: gI- 31(35%) children with an unfavorable course of HCM [death n = 6, CA n = 4, 5pts were qualified for HTx (performed in 1pt, 1pt is waiting for HTx, 3pts died on waiting list), 9pts required surgical myectomy, in 12pts ICD was implanted (4 pts as a secondary, 8 pts as a primary prevention)] and gII- 57pts (65%) who were stable, treated only pharmacologically. Patients demographics, clinical symptoms, family history of SCD as well as the results of echocardiography, 12-leads ECG, 24-h Holter ECG were analyzed and compared between the groups.

**Results:** Children in both groups do not differ regarding age at diagnosis, incidence of syncopes, chest pain, family history of SCD, however pts in gI more often had symptoms such as fatigability, exertional dyspnea (39% vs 11%;  $p = 0.017$ ). Patients in both groups differed significantly regarding NYHA functional class ( $p = 0.01$ ). In gI septal thickness (333% vs 220% of mean normal range relative to BSA;  $p < 0.0001$ ), thickness of the posterior wall of LV. (179% vs 138%;  $p = 0.0037$ ) were significantly higher and more frequently LVOTO. (48% vs 16% pts;  $p = 0.001$ ) was observed. The Sokolow-Lyon index (66 mm vs 44 mm;  $p = 0.0004$ ), QTc interval (435 ms vs 409 ms;  $p = 0.002$ ), QTc dispersion (52 ms vs 44 ms;  $p = 0.047$ ) were significantly higher and nsVT was more frequent (24% vs 5% pts;  $p = 0.009$ ) in gI.

During follow up, 9pts in gI died, 6(6,8%) from progressive heart failure and 3 (3,4%) from SCD, mean annual mortality rate was 1,48%.

**Conclusions:** 1. There was a significant correlation between LV hypertrophy, LVOTO and adverse clinical course of HCM and the need for more aggressive therapy. 2. The value of the Sokolow-Lyon index, QTc interval, QTc dispersion and the presence of nsVT have been associated with a worse prognosis in children with HCM.

**P-140****QTc interval in patients with non-compaction of the myocardium**

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Non-compaction of the myocardium is a genetically determined cardiomyopathy with wide range of outcomes. The aim of this study was to assess the association of non-compacted myocardium and prolonged QT interval in children.

**Material and methods:** Patients less than 18 years of age with diagnosis of isolated non-compaction of the myocardium were reviewed for clinical, ECG and echocardiographic data and outcomes. QTc interval was measured according to the Bazett formula.

**Results:** From 1996 to 2010, 43 patients (23 males), aged 0 to 217 months at diagnosis (median 9.3 months), were followed-up for 0.5 to 99 months (median 8.2 months). ECG was abnormal in all cases, QTc interval >440 ms in 23% (QTL = 10 cases), <440 ms in 77% (QTN: 33 cases). Twenty-two presented with heart failure (51%), 2 with shock and cardiac arrest, 3 with arrhythmia, 2 with syncope or chest pain and 14 had no symptom (32.5%), with no difference between QTL and QTN cases. Familial recurrence was 21% (more frequent in QTL: 40% vs 15%) and parents consanguinity 14% (50% if QTL vs 19% in QTN). N/C index was  $2.5 \pm 0.7$  (median 2.3): mean 3.1 in QTL group and 2.2 in QTN group ( $p = 0.01$ ). LV apex was involved in 96.7%, with more than 3 locations in 25.5% (33.3% in QTL, 22% in QTN) and RV involvement in 9.3%. Shortening fraction was  $25.4 \pm 11.6\%$  (median 25%), not different with QTc. Nine patients were diagnosed with previous small muscular VSD (none ranged in group QTL). Six patients died (13.9%) at median age 6.2 months (3.6 to 43) and median follow-up 2.7 months (0.5 to 12.5). Survival was not different between QTL and QTN patients. Five underwent heart transplantation (11.6%) at median follow-up 2.8 months (1.6 to 27.4). At least one episode of heart failure occurred in 18 (42%), arrhythmic or thromboembolic events in respectively 5 (11.6%) and 3 (6.9%). Bad events were more frequent in QTL group (80%) than in QTN. (57.5%,  $p = 0.1$ ).

**Conclusion:** This study showed a specific association of non-compaction cardiomyopathy with prolonged QT interval in children, which is correlated with familial occurrence, N/C index and poor outcomes.

**P-141****Population study of 332 consecutive newborns with HLHS – a single center experience**

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**Background:** Polish Mother's Memorial Hospital, Research Institute is one of the leading centers for hypoplastic left heart syndrome (HLHS) treatment in Poland.

**Aim:** The aim of the study was the estimation of perinatal and anatomic data of 332 newborns with HLHS operated at our institution.

**Material and Methods:** Retrospective analysis of perinatal data and anatomical findings of 332 newborns with HLHS. (226 boys – 68% and 106 girls – 32%) treated by staged Norwood operation at our institution in 1992–2010.

**Results:** Prenatal diagnosis was made in 58% of pts (68–75% in last 5 years), mean pregnancy duration was  $39.4 \pm 1.8$  (31–43) weeks; 77.3% of pregnancies finished by vaginal births, 22.7% by cesarean section, mean mother's age was  $26 \pm 5$  (17–43) years, in 147 cases (44%) child with HLHS came from the first pregnancy. Mean Apgar score was  $8.7 \pm 1.4$  (1–10), mean birth mass was  $3236 \pm 505.6$  g (1995–4430 g). 10 patients (3%) came from twin pregnancies (in 1 case the second twin with TOF, in remaining 9 – the second twin was healthy). 5 patients (1.5%) had coexistent extracardiac malformations. In 4 cases (1.2%) our patient was the second child with HLHS of the same parents. In 6 families (1.8%) cardiac or extracardiac malformations in HLHS patient's siblings were confirmed (in 2 families TGA in sibling). In one patient Turner syndrome (45,X) was diagnosed.

Anatomic subtypes of HLHS: MA/AA in 123 pts (37%); MS/AA in 113 pts (34%); MS/AS in 93 pts (28%); MA/AS in 3 pts (1%). Myocardial performance index for right ventricle (RV-MPI)  $0.521 \pm 0.18$  (0.2–0.968) vs  $0.3 \pm 0.078$  (0.183–0.445) in control group of 50 healthy newborns. Restrictive atrial communication was confirmed in 33 pts (10%). Mean ascending aorta diameter was 3.8 mm (1–7.5 mm). Severe tricuspid regurgitation was diagnosed in 40 patients (12%).

**Conclusions:** Patient with HLHS is usually male, good developed, full term delivery newborn of young mother. HLHS rarely coexists with other malformations or genetic disorders. In our material MA/AA, MS/AA, MS/AS subtypes occurred with similar frequency, MA/AS was very rare. RV-MPI for HLHS patients is significantly higher comparing with healthy neonates.

**P-142****Intima-media thickness measurements in children after end-to-end operation of coarctation of the aorta**

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The measurements of carotid intima-media thickness (cIMT) is a good predictor of arterial wall damage, atherosclerosis, in adults with arterial hypertension, recommended by European Society of Hypertension. Data on cIMT in children after end-to-end operation of coarctation of the aorta (CoA) are not available.

**Aims:** to check the relations between the cIMT and the time of surgical repair, the duration of follow-up, and arterial hypertension in children after end-to-end operation of CoA.

**Material and Methods:** In 50 pts operated at the age below 3 mths (I group – 15 pts) and after 3 mths (II group – 35 pts) ABPM and cIMT measurements were performed  $8.82 \pm 1.94$  and  $10.71 \pm 1.96$  years after end-to-end operation of CoA. The groups with (HT+) and without (HT-) arterial hypertension were established on the base of ABPM results. The mean age of pts at the time of first and second assessment were  $10.14 \pm 3.57$  and  $12.18 \pm 3.53$  yrs.

**Results:** Arterial hypertension was found in 35% of pts (20% of I group and 43% of II group). Increased right cIMT (with reference to norms for age and sex) was found in 35% of pts, left cIMT in 33% of pts. cIMT was slightly increased in group II and HT+.

There was a moderate correlation between the mean value of right arm systolic blood pressure and right cIMT ( $r < 0.36$ ,  $p < 0.02$ ) and moderate correlation between age at operation and right and left cIMT ( $r < 0.44$ ,  $p < 0.01$  and  $r < 0.43$ ,  $p < 0.01$ ). There were no statistically significant difference in right and left cIMT between two examinations (the study was repeated after  $1.97 \pm 0.28$  yrs).



*Conclusions:*

- In 33% of patients after end-to-end operation for coarctation of the aorta carotid IMT is increased in long follow-up.
- There are significant, positive correlations between carotid IMT and right arm systolic blood pressure, the age at surgery and the duration of follow-up.
- It seems to necessary to examine a more patients for assessment a importance of measurements of IMT in patients after operation of coarctation of the aorta.

**P-143****Hypoxic myocardial injury in newborn infant-diagnosis and evolution**

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*Purpose:* To present the main aspects of perinatal myocardial injury and the utility of echocardiography for its diagnosis and follow up. *Methods.* Patients: 82 newborns aged 0 to 14 days, with normal birth weight, with perinatal hypoxia (Apgar score 3–7), receiving resuscitation, but without major congenital heart diseases. All cases were investigated by clinical exam, ECG, chest X-ray (Rx.CT), Doppler echocardiography (Echo). Could not be investigated some cardiac biomarkers as CPK-MG and cardiac troponin. Most of patients were evaluated clinically and echo after 6 months. *Results.* The patients had mainly signs of neurological post hypoxic suffering, only 6 cases signs of severe heart suffering (cardiomegaly, respiratory distress, cyanosis, peripheral circulatory failure), otherwise the cardiac exam revealed: systolic murmur (64 cases) and signs of persistent pulmonary hypertension of newborn (PPHN) 7 cases. Chest X-ray: cardiomegaly (31 cases–37,8%). ECG: severe left ventricle (LV) repolarization disturbances and low voltage of QRS complexes (37), without ischemic changes. Doppler echo exam at 2–7 days of life revealed: \* the absence of other severe congenital cardiac anomaly; \* permeability of foramen ovalae (100%) and forced foramen ovalae (gradient LA/RA > 8 mmHg); mild to severe tricuspid insufficiency and RV and RA dilation (29); sometimes right-left shunt through the FO \*myocardial hypertrophy (42 cases) mainly IVS. (29), signs of PPH. (6); prolonged IVRT. (35), increased myocardial performance index (44 cases), the systolic dysfunction in 5 cases and severe LV diastolic dysfunction in 45 cases. All the cases received spironolactone 1–2 mg/kg/day for 3 months. Reevaluation at 6 months showed the reduction of the myocardial hypertrophy and of tricuspid regurgitation, with a normal LV systolic and diastolic function. *Conclusions.* The perinatal hypoxia can induce a important myocardial injury as hypoxic ischemic myocardopathy or transient post hypoxic hypertrophic cardiomyopathy at more than 62,2% of patients, the signs of cardiovascular suffering missing often. Echo is the main method for diagnosis and follow up of perinatal hypoxic cardiomyopathy and is necessary performed from the first week of life. The research of cardiac biomarkers CPK-MB and cardiac troponin may amplify the value of cardiological investigation of hypoxic myocardial injury in newborn infant.

**P-144****Heart disease in the newborn of diabetic mother. Usefulness of echocardiographic assessment**

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*Objectives:* To present the main aspects of cardiac involvement in newborn of diabetic mother and utility of echocardiography (Echo) for the diagnosis and follow-up their evolution.

*Methods:* Patients: 76 newborns of diabetic mothers, follow up over a period of 6 years. Patients were investigated in the first week of life and 49 were re evaluated at 6–12 month, by clinical exam, ECG, cardiothoracic radiography (Rx.CT) and Doppler echo for congenital and/or acquired cardiac diseases. Fetal echo was performed in 36 cases after 28 weeks of gestation.

*Results:* Fetal echo showed: cardiomegaly and myocardial hypertrophy of left ventricle (LV) in 9 cases, confirmed by postnatal echo. Clinical exam in newborn: macrosomia in 40% cases, in 32 patients a systolic murmur was found, only 2 cases with signs of heart failure and the other being asymptomatic or presenting signs for other pathology than cardiac. ECG: LV hypertrophy in 14 cases and disturbed ventricular repolarization in 30 cases. Rx.CT: cardiomegaly (12). Echo showed: non obstructive hypertrophic cardiomyopathy (HCMP) with asymmetric IVS hypertrophy (32 cases: 42%), arterial pulmonary hypertension (5), LV diastolic dysfunction with normal systolic function (52% of cases) and congenital cardiac anomalies: PDA. (6), VSD. (3), coarctation of aorta (1), ASD. (4 cases). LV myocardial hypertrophy was not significantly correlated with the type of mother's diabetes, before pregnancy or gestational, but rather to an inadequate control of disease. Control performed at 6–12 months (21 cases) revealed a normal morphological cardiac aspect in 14 cases or significant reduction of HCMP. (7 cases), all of them showing normal diastolic and systolic LV function.

*Conclusions:* Newborn of diabetic mother presents a high risk for cardiac involvement, either cardiac congenital malformations (19%) or acquired cardiac pathology: HCMP. (42% of cases) and disturbances of diastolic function of LV. (53%). This fact justify early cardiologic screening for all of these newborns with or without of cardiac suffering symptoms. Fetal echo provides useful data for diabetic pregnant women and should be made mandatory to all these patients. Echocardiography is the most sensitive and noninvasive method of diagnostic, useful for primary diagnostic as well as for follow up.

**P-145****Association of mitral regurgitation with anomalous left coronary artery from the pulmonary artery (ALCAPA) in children**

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*Objective:* To evaluate the frequency and association of isolated mitral regurgitation with ALCAPA syndrome in children presenting with left ventricular dysfunction or dilated left ventricle.

*Methods:* 20 year review of ALCAPA syndrome from two paediatric cardiac centres in United Kingdom was undertaken. Patients with abnormal coronary artery origins apart from ALCAPA syndrome were excluded.

*Results:* Eighteen consecutive patients were identified. Twelve of them were females (66%). Age at presentation varied from one month to seventeen years. The age from first presentation to suspect the diagnosis of ALCAPA varied from one week to eight years. Only 3 patients had the diagnosis made at presentation (16%). The commonest reasons for referral were features of cardiac failure in 9 (50%), and respiratory distress or asthma in 8 (44%) patients. 5 (27%) patients were investigated for failure to thrive. The common initial diagnosis was dilated cardiomyopathy



in 12 (66%). The echocardiographic features at presentation included LV dysfunction in 11 (61%), significant mitral regurgitation in 8 (44%). Mitral regurgitation was considered as the primary pathology in 8 patients (44%) hence its association with ALCAPA was overlooked for as long as 5 years. On the contrary of general view, with ultrasound examination only 4 (22%) patients exhibited dilated right coronary artery, and in 5 (27%) the origin of left coronary artery could not be clearly visualised. 8 (44%) showed abnormal retrograde flow in the pulmonary artery. The commonest ECG findings were non-specific ST segment changes in 11 (61%), pathological Q wave in 14 (77%) in lead aVL, and T wave inversion in 10 (55%) in leads aVL and V6. 8 (44%) patients underwent cardiac catheterization to confirm the diagnosis. All 18 patients had surgery immediately after diagnosis. Post-surgical follow up echocardiogram showed resolution of LV dysfunction in 10 (55%) patients. 3 (16%) patients died following surgery.

**Conclusion:** Isolated mitral regurgitation is an important and frequent finding in children with ALCAPA syndrome. Any child with echocardiographic diagnosis of isolated significant mitral regurgitation and left ventricular dysfunction or dilatation should prompt a search for ALCAPA syndrome.

#### P-146

##### Long-term outcome of ALCAPA. A single centre experience

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**Objectives:** To evaluate a single centre's experience of the management and long term outcome of anomalous origin of left coronary artery from pulmonary artery (ALCAPA).

**Methods:** Retrospective analysis of the case notes from September 1990 to October 2010.

**Results:** Total 25 patients (6 male, 19 female) had a diagnosis of ALCAPA with following demographics

	Median	Range	
Age at repair	5.50	(1.80–101.90)	Months
Weight	5.81	(3.99–30.80)	Kg
Bypass time	93.00	(38–351)	Minutes
Cross clamp	48.50	(33–97)	Minutes
Hospital stay	12.50	(6–87)	Days

At the time of presentation, 20 patients had moderate to severe impairment of left ventricular function with a median fractional shortening (FS) of 12% (range 5%–32%) and 15 patients had moderate to severe mitral regurgitation. Surgery was performed immediately on diagnosis with coronary re-implantation in 15 patients and Takeuchi repair in 9 patients and 4 patients needing mitral valve repair.

Long term follow up data is available for 20 patients. The median duration of postoperative follow up was 72 (range 0.5 to 214) months. All 20/20 patients are alive with 100% survival rate. At last follow up, 19/20 patients were asymptomatic, the left ventricular function was normal in 17 patients, mildly impaired in 2 patients and severely impaired in 1 patient (median FS of 35% (range of 23% to 43%)). Significant mitral regurgitation was present in only 2 patients. One patient who had Takeuchi tunnel

had balloon dilation of pulmonary trunk, 2 patients with Takeuchi tunnel and one patient in direct implantation group had restenosis at the origin of left coronary artery which were dealt surgically.

**Conclusion:** Surgical repair of ALCAPA has got good long-term results with low rate of mortality and re-intervention. The ventricular dysfunction and mitral regurgitation usually gets better with reperfusion.

#### P-147

##### ST-segment elevation myocardial infarction (STEMI) due to severe anemia resulting from transient erythroblastopenia of childhood in a 2 year old boy after arterial switch operation in the absence of coronary artery stenosis

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**Background:** Myocardial ischemia or infarction occurs when the oxygen demand of the myocardium exceeds its supply. Severe anemia reduces the oxygen supply of the myocardium but increases the cardiac output and may therefore cause an oxygen supply-demand mismatch. A myocardial infarction due to severe anemia in children is extremely rare.

**Case report:** Arterial switch operation (ASO) was performed on a 9-days-old boy with transposition of the great arteries. The postoperative course was uneventful except for a moderate supravalvular pulmonary stenosis and consecutive right ventricular hypertrophy. Aged 1 year 9 months he presented with a history of fatigue and pallor of several days. Diagnostics revealed a severe normochromic normocytic anemia and an acute myocardial infarction. Hemoglobin was 1,7 mmol/l = 2,7g/dl, hematocrit was 8%, cardiac troponin I was max. 0,98 ng/ml (normal <0,04 ng/ml) and CK-MB was max. 10,5 ng/ml (normal <6,6 ng/ml). The electrocardiogram showed ST elevation in leads aVR and V1 and ST depression in leads I, II and V4, V5 and V6. A right ventricular infarction was diagnosed. The patient responded well to erythrocyte transfusion with normalization of laboratory findings and physical condition. The recovery was uneventful. Further diagnostics suggested transient erythroblastopenia of childhood to be the cause of the anemia. Coronary angiography was normal.

**Discussion:** This 2-year-old patient after ASO suffered a ST-segment elevation myocardial infarction (STEMI) due to severe anemia resulting from transient erythroblastopenia of childhood. Coronary artery stenosis as a possible cause of myocardial ischemia after ASO was ruled out. To our knowledge no similar case of myocardial infarction due to anemia has been described in children so far.

#### P-148

##### The Namibian Children's Heart Project: A new cardiac service in Africa

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**Introduction:** Until recently there were no services for Namibian children or adults with heart disease. The aim of this project, initiated by the Ministry of Health, is to develop a self sufficient, sustainable cardiac service for children and adults in this country. The Cardiac Unit was commissioned in Windhoek in October 2010 and this paper describes our 2 year experience.

**Methods:** This prospective study was conducted between September 2008 and December 2010 at Windhoek Central, Christiaan Barnard Memorial and Panorama Medi-Clinic Hospitals. Patients were recruited from the Paediatric and Congenital Heart Disease and Rheumatic Heart Disease Clinics. Funding was initially provided by businessman and philanthropist Harold Pupkewitz and thereafter, by the Namibian Government.

**Results:** During the 2.3 year period 71 indigent patients were referred to Cape Town for surgery or intervention. A further 20 received surgery at Windhoek Central Hospital. The age range was 3 months to 33 years, mean 8 years. Fourteen were over 18 years. Cardiac catheterization demonstrated 3 patients inoperable and 1 did not require intervention or surgery. Two patients were referred back to Windhoek for further investigation of non-cardiac disease. Of 85 patients aged 3 months to 29 years, 8 had interventional procedures and 76 heart surgery. Fourteen patients had rheumatic heart disease and 3 adults had congenital heart disease. Tetralogy of Fallot (19), ventricular septal defect (17), patent ductus arteriosus (13) and atrial septal defects (5) were the commonest congenital heart diseases. There were 2 early deaths (operative mortality 2%) and 2 late deaths. There were 8 re-operations in 6 patients. Three (3) patients needed surgery for post-operative complications.

**Conclusions:** The absence of curative heart services for children has led to an older demographic with high numbers of adult survivors with congenital heart disease. It also means low numbers of children seen with critical congenital disease. This project demonstrates that goodwill, political commitment, regional cooperation and public private partnerships can together bring great benefit to indigent, previously neglected children. Furthermore they can lead to the development of self sufficient service with thus far, satisfactory results.

#### P-149

##### **Ebstein's anomaly: Rhythm disturbances after modified reconstruction of tricuspid valve without ventricle plication**

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**Introduction:** Ebstein's anomaly (EA) is a rare congenital heart defect of the tricuspid valve in which the hinges of the septal and/or posterior leaflets are displaced downward to the right ventricle. The anterior leaflet is usually not displaced but is enlarged and sail-like and valve closure is likewise displaced downwards.

**Methods:** Since 1988 we have operated patients with Ebstein's anomaly (EA) using a modified repair technique of the tricuspid valve. This technique reconstructs the valve mechanism at the level of the true annulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber. Additional attachment of the anterior right ventricle wall to the interventricular septum ("Sebening stich") and if possible, reconstruction of a tricuspid valve as a double orifice valve was performed in a modified technique since 2004.

Preoperative, perioperative and postoperative arrhythmias in 57 consecutive patients who underwent operation for EA were reviewed. There were 22 male and 35 female patients (median age 22 years, range 3 months to 68 years. Median follow-up period was 33 months (range 1 month to 22 years).

**Results:** Early mortality was 7.1%, late mortality 5.2%, since 2004 no patient died. Re-operation was necessary in 2.3%. Preoperative rhythm disturbances were present in 54% of all

patients [n = 4 with WPW-syndrome, n = 6 with bradycardia and n = 21 with supraventricular tachycardia (SVT)]. 35% had intraventricular conduction system disturbances [n = 13 with incomplete right bundle branch block (RBBB), n = 7 with complete RBBB and no complete atrioventricular block].

Perioperative SVT was reduced from 37% to 12%, and late follow-up demonstrated an incidence of 14% for SVT. Bradycardia was reduced from 11% preoperatively to 3.5% in the late follow-up. No WPW syndrome was documented after the operation. Postoperative complete RBBB increased from 12% to 32% and complete atrioventricular block was observed in 22%. Analysis of the type of severity of EA (due to the Carpentier classification) seems to demonstrate increasing rhythm disturbances in more severe forms of EA.

**Conclusions:** Rhythm disturbances in patients with EA are common. Reconstruction of the tricuspid valve even without ventricle plication seems to minimize the incidence of supraventricular tachycardia, the incidence of postoperative complete atrioventricular block was significant.

#### P-150

##### **Changing Trends in GUCH Intervention over last 10 years. A single Centre experience**

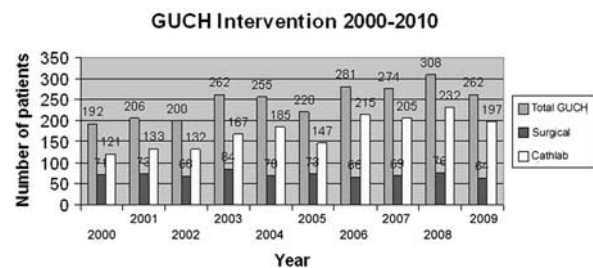
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**Introduction:** Percutaneous interventional approaches are increasingly recognised as a preferred treatment option Adult with Congenital Heart Disease (GUCH). The changing management of the GUCH population is important, particularly with regards to service provision, so we assessed the GUCH population undergoing interventional catheterisation in our institution over the last 10 years, assessing number and types of intervention and comparing to those undergoing surgery.

**Methods:** A database of all patients undergoing cardiac catheterisation at our Centre was assessed, examining the number and types of catheter procedure performed in patients over the age of 18. This was then compared to a similar surgical database.

**Results:** Over the ten year period 2327 patients underwent a cardiac catheter or surgical intervention—711 surgery (30.6%), 1616 (69.4%) a catheter based intervention. An additional 379 had a diagnostic catheter procedure. From 2000–2004, 983 patients underwent a procedure, followed by 1344 in the period 2005–2009. The number and type of intervention (surgical vs percutaneous) is shown in Figure 1.



In the first 5 years (to the end of 2004), 752 patients underwent a cardiac catheterization with 620 (82.4%) having an intervention. In the last five years (2005–2009), 1242 patients underwent catheterization with 996 (80.2%) having an intervention.

**Conclusions:** In our Centre increasing numbers of adults are having percutaneous congenital/structural interventions. The number having surgery has remained constant. Most intervention has been of interatrial defects with an increase in those having PFO closure and decrease in those having ASD closure. The actual numbers of those having coarctation stenting, ventricular septal defect closure and PDA closure have remained roughly similar but there has been an increase in the number having percutaneous intervention to the right ventricular outflow tract. The changing interaction of percutaneous intervention and surgery in the GUCH population continues to evolve and needs study to provide optimal service levels for the GUCH population.

#### P-151

##### Fallot repair in adulthood. Surgical results and medium follow-up

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**Objectives:** Evaluate the surgical risk and late benefits in Fallot patients repaired at age >18 years.

**Material and Methods:** 27 patients operated in 2 hospitals, between 1993 and 2010. Retrospective study of the patient's clinical reports. The statistical analysis was done with SPSS-15.0.

**Results:** 55% males, 30% right aortic arch, 48% had ASD. The diagnoses were PE + VSD: 3; DCRV + VSD: 10; Fallot: 11, DORV Fallot type: 2; PA + VSD: 1. Prior palliative surgery 26%. Preoperative NYHA status  $\geq$ III 59%. Preoperative arrhythmias 26%. SaO<sub>2</sub>: 91%  $\pm$  7; hematocrit 50  $\pm$  11. Ten patients were cyanotic.

RV-PA preoperative ECO gradient: 95 mmHg  $\pm$  34. PR and TR is absent or mild in 96%. RV function: normal 74%, mild dysfunction 15%, moderate 7%, severe 4%. Catheterization in 23 patients: RV-PA gradient 81  $\pm$  33 mmHg. Mean systolic and diastolic RV pressure were 116/12 mmHg; systolic pulmonary pressure 33  $\pm$  15 mmHg. Cardio MR in 10 patients.

Indication for surgery: in asymptomatic patients peak/mean ECO gradients > 60/40 mmHg and in symptomatic >50/30. CPB and aortic cross-clamp time were 121  $\pm$  40 and 88  $\pm$  39 minutes. The VSD was closed in 52% via RA, 41% via RV and 7% via the aorta. Pulmonary annulus was preserved in 68%, pulmonary bioprosthesis 18%, 11% transannular patch. Associated surgery: 2 aortic prostheses and 3 valvuloplasty (1 aortic, 1 mitral, 1 tricuspid)

No hospital mortality. Intubation time: 16 hours  $\pm$  49, intensive care stay 3  $\pm$  2 days, hospital stay 14  $\pm$  7 days. Mean follow-up was 7 years (0.5 to 17). There has not been late mortality. One patient required interventional catheterization for stenting RPA and 3 needed reoperation: residual VSD, pulmonary bioprosthesis+residual VSD and tricuspid bioprosthesis.

Actual functional class is  $\leq$ II: 96%; 52% have no medical treatment. PA-RV ECO gradient: 21  $\pm$  14 mmHg.

**Conclusions:** Fallot repair in adulthood is performed in our hospitals without hospital mortality. The long-term survival is excellent, with 11% reoperation in late follow-up. In DORV Fallot type and TOF the percentage of pulmonary annulus conservation is superior to the children's series. Pulmonary bioprosthesis improve postoperative course when transannular surgery is required

#### P-152

##### Adult life reoperations in Fallot patients with corrective surgery during childhood

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**Objectives:** In our Grown up Congenital Heart (GUCH) Unit are currently followed 264 patients with Fallot corrective surgery during childhood. We analyze late reoperations during adulthood in this group and present the long term evolution of these patients.

**Material and methods:** Retrospective study from clinical reports of 48 reoperations in 46 patients. Statistical analysis was done with SPSS-15.0.

**Results:** Mean age: 30  $\pm$  9 years, 71% males, number of previous surgeries per patient: 2  $\pm$  1. One patient has only palliation (2%), 52% have only correction, and 46% have both surgeries (palliative + corrective). Mean time interval between the corrective surgery and reoperation was 22  $\pm$  9 years. Preoperative functional NYHA class was II. (38%), III-IV. (54%). Arrhythmia was present in 51%. Main indications for surgery were: pulmonary insufficiency (37%), pulmonary stenosis (19%), double pulmonary lesion (17%), aortic insufficiency (13%), residual ventricular septal defect (8%), and tricuspid insufficiency (6%).

Reoperations were done with extracorporeal circulation and moderate hypothermia, femoral cannulation was used in 59%, aorta was not clamped in 29%. Attending to the surgical techniques used: 69% patients received a pulmonary prostheses, 46% underwent septal defect closure, 23% had surgery on the tricuspid valve, 21% received an aortic prostheses, and 10% had a maze procedure. Mean postoperative intubation time was 15  $\pm$  22 hours, intensive care unit stay was 3  $\pm$  2 days, and Hospital stay was 12  $\pm$  8 days. Hospital mortality was 6,3% (3 patients).

Mean follow up after the reoperations was 4,3  $\pm$  4,6 years. During this period, 2 patients (4,4%) had late mortality, 2 patients needed percutaneous angioplasty (4,4%), and 3 patients (6,6%) needed other surgical reintervention. Actually 59% patients are in NYHA functional class I, 31% in class II, and 10% in class III.

**Conclusions:** Fallot corrective surgery during childhood shows good prognosis in the long term, but some patients (18%) will need reoperations along their adult lives. Principal indications for reintervention are pulmonary regurgitation and/or stenosis needing the interposition of a bioprosthesis. Reoperations in these patients in our Unit can be achieved with good results and low mortality at short and long term.

#### P-153

##### Influence of pulmonary valvulation in adults with Tetralogy of Fallot on left ventricular function

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The impact of pulmonary valvulation in adults with repaired Tetralogy of Fallot and significant pulmonary regurgitation on RV volume and function has widely been studied. However its influence on LV function is less clear. The aim of this study is to compare LV function prior and after pulmonary valvulation.

**Material and Methods:** Clinical status, physical ability (Max vO<sub>2</sub>), ECG and 24h-ECG, echocardiography data including RV and LV diameters and systolic function, RMN when available, were analyzed before and after pulmonary valvulation in adults with surgically corrected Tetralogy of Fallot.

**Results:** 49 GUCH patients, who underwent repair of Tetralogy of Fallot from 1970 to 1999, were included in the study: 29 needed a previous palliative shunt, 38 had an infundibulo-transannular patch. Pulmonary valvulation was performed at a mean of 24 years after corrective surgery (mean age at initial surgery and at valvulation respectively: 5.5 and 29.4 y). Mean follow-up after valvulation was 1.5 y. No patient was lost to follow-up, no death occurred. Although NYHA status improved (from 1.9 to 1.1), physical ability did not change with maximal power level of 120 Watts before and 125 Watts after valvulation (NS) and max VO<sub>2</sub> 20.8 versus 21.5 ml/min/kg (NS). ECG showed no difference in QRS duration (158 vs 155 ms), but 24h-ECG showed decrease in significant arrhythmias (atrial fibrillation or flutter, polymorph ventricular premature beats or ventricular tachycardia). Echocardiographic records showed decreased RVEDD. (41 vs 29 mm,  $p < 0.0001$ ), increased LVEDD. (43 vs 47 mm,  $p = 0.001$ ) and increased LV shortening fraction (31 vs 36%,  $p = 0.01$ ). RMN analysis similarly demonstrated decreased RV volumes (diastolic 172 vs 94 ml/m<sup>2</sup>,  $p < 0.0001$  and systolic 113 vs 62 ml/m<sup>2</sup>,  $p = 0.01$ ) but unchanged RV ejection fraction, and increased LV diastolic volume (58 vs 67 ml/m<sup>2</sup>,  $p = 0.02$ ) and LV ejection fraction (55 vs 61%,  $p = 0.04$ ).

**Conclusion:** Pulmonary valvulation in Fallot patients with significant pulmonary regurgitation does not yield to improve objective physical ability, QRS duration, RV function, but contributes to ameliorate subjective physical status and has a favorable impact on RV volumes, LV function, and the occurrence of severe arrhythmias.

#### P-154

##### **Pregnancy: tolerance and outcomes in women with Ebstein's anomaly**

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Pregnancy in women with congenital heart disease is a challenge for cardiologists and obstetricians, facing the potential risk of serious events. In women with Ebstein's anomaly, pregnancy may induce arrhythmias or increase cyanosis and dyspnea, and therefore has long been contraindicated.

**Aim:** To assess the tolerance and mother and offspring outcomes in pregnant women with Ebstein's anomaly, in an attempt to define some factors of risk.

**Material and methods:** clinical status and outcomes of pregnancy (mother and offspring) in women with Ebstein's anomaly were analyzed. The presence of a Wolf-Parkinson-White syndrome and/or cyanosis was recorded.

**Results:** 22 women had 47 pregnancies (1 to 6 pregnancies; 2.13 pregnancies/woman): 4 were cyanotic and had 6 pregnancies (1.5/w) with 5 livebirths and 1 therapeutic abortion. Premature birth occurred in 3 of the 5 livebirths (60%). The 18 non cyanotic women had 41 pregnancies (2.3/w) with 3 miscarriages, 1 abortion and 37 live births (90%). Prematurity occurred in 7/37 live births (19%). Mean birth weight was respectively 2.130 and 2.660 kg for the newborns from cyanotic and noncyanotic mothers ( $p = 0.03$ ). All pregnancies were well tolerated, no maternal death occurred. 11 women out of 22 (10/18 non cyanotic vs 1/4 cyanotic) had no cardiac symptom during pregnancy. 5 of them were in Stade A of Carpentier, 4 in stade B and 2 in grade C. Dyspnea increased in 3 of 18 noncyanotic vs 3 of 4 cyanotic, asthenia in 3 of 18 vs 3 of 4 ( $p = 0.02$ ). 7 women

had a Wolf-Parkinson-White syndrome (32%): of them 4 (57%) had no arrhythmia, 2 experienced palpitations and 1 had a well tolerated supra-ventricular tachycardia (SVT). Among the 15 women without WPW syndrome, 10 (67%) had no arrhythmia, 3 had palpitations and 2 SVT. Neither life-threatening nor severe arrhythmia occurred

**Conclusion:** Pregnancy in women with Ebstein's anomaly is well tolerated, especially in non cyanotic mothers (less arrhythmias, less prematurity, greater birth weight). Therefore, Ebstein should not be a contra-indication for pregnancy, even in cyanotic women. However, to insure optimal tolerance, preventive closure of the inter-atrial shunt might be proposed prior to pregnancy.

#### P-155

##### **Exercise capacity and self reported functional status in patients with complex congenital heart disease**

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**Objectives:** To compare self reported quality of life with measured exercise capacity in patients with complex congenital heart disease.

**Methods:** Forty-eight patients (20 female, age 18–55years old) with various congenital heart defects (19 TOF/DORV, 16 TGA, 6 Fontan, 2 PAH, and 5 others) in NYHA I or II functional class completed an SF36–health related quality of life questionnaire and performed a six-minute walk test (6MWD), followed by a cardiopulmonary exercise test (predicted MVO<sub>2</sub>).

**Results:** Majority of patients in NYHA class I achieved a 6MWD of 450m (97%) and reported a physical functioning score and a mean QOL index above average (88% and 87%, respectively), however only 38% of them had an MVO<sub>2</sub> >70% predicted. Patients with NYHA class II were less likely to have an MVO<sub>2</sub> >70% predicted (7%), but 43% of them achieved a 6MWD of 450m and more than half of them reported a physical functioning score and mean QOL index above average (71% and 58%, respectively). Self reported physical functioning score and average QOL index correlates significantly with 6MWD ( $r = 0,501$ ;  $p < 0,001$  and  $r = 0,602$ ;  $p < 0,001$ , respectively), but not with predicted MVO<sub>2</sub>.

**Conclusion:** Patients with complex congenital heart disease tend to overestimate their functional status in spite of a decreased exercise capacity. The most objective measurement of a decrease in exercise capacity seems to be the predicted peak oxygen uptake.

#### P-156

##### **The clinical implications of serum uric acid levels in adult patients with congenital heart disease**

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**Background and aims:** Serum uric acid (UA) is a strong predictor of cardiovascular risks in patients with chronic heart failure. In adult patients with congenital heart disease (ACHD), the prevalence of hyperuricemia is largely unknown. It also remains undetermined whether hyperuricemia can predict cardiovascular events in ACHD. The aim of this study was to investigate the prevalence of hyperuricemia and to assess the clinical contribution of serum UA to the risk of cardiovascular events in ACHD.

**Methods and Results:** A total of 257 patients were enrolled (mean age 30 +/- 10 years, 45% female). The mean UA level in this



cohort was  $5.8 \pm 1.6$  mg/dl and 46 patients (18%) had hyperuricemia. During a follow-up of  $42 \pm 19$  months, 66 patients (26%) experienced a cardiovascular event (cardiac death, symptomatic arrhythmia, hospitalization due to worsening heart failure, or thromboembolism). Patients with hyperuricemia had higher risk for cardiovascular events compared with patients without hyperuricemia, (46% vs 22%,  $p < 0.05$ ). On multivariate regression analysis, a high serum uric acid level was associated with the hemoglobin level ( $p = 0.015$ ), serum creatinin level ( $p = 0.005$ ) and aortic desaturation ( $p < 0.001$ ). The independent predictors of hyperuricemia were found to be male gender (hazard ratio 2.58,  $p = 0.003$ ), diuretics use (HR 2.52,  $p = 0.004$ ), hemoglobin level (hazard ratio 2.61,  $p = 0.007$ ), and aortic desaturation (hazard ratio 1.42,  $p = 0.025$ ). Kaplan-Meier analysis indicated that patients with hyperuricemia had a higher incidence of cardiovascular events. However, Cox proportional hazard analysis showed that hyperuricemia was not an independent predictor of cardiovascular events, while diuretics use was a strong predictor.

**Conclusions:** Hyperuricemia is relatively common in ACHD. High serum uric acid levels are associated with polycythemia, impaired renal function, and aortic desaturation. Although hyperuricemia may be a predictor of morbidity and mortality in ACHD, it is not independent of several confounders, especially diuretic use.

#### P-157

##### **Clinical outcome and echographic features of patients with repaired tetralogy of Fallot and biventricular pacing**

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**Introduction:** Right heart failure is a common feature in patients with repaired tetralogy of Fallot (TOF), right ventricular (RV) dysfunction and right bundle branch block (RBBB). Biventricular pacing (BVP) has been described as a potentially useful therapeutic in some cases. We aimed to investigate the clinical outcome and dyssynchrony echocardiographic characteristics of patients with repaired TOF and BVP.

**Methods:** A systematic retrospective study of all of the patients of CHU de Bordeaux with repaired TOF and BVP was realized. Clinical NYHA status and exercise test performance were retrieved before and 6 months after BVP. All patients benefited from an echocardiography with dyssynchrony measures in spontaneous rhythm, RV pacing and BVP.

**Results:** 10 patients (7 male,  $36.6 \pm 13$  years old) were retrieved from our database. Surgical repair had occurred at the age of  $7.4 \pm 5.8$  years. BVP was effective since  $18 \pm 10$  months.

After 6 months of BVP were noted a significant improvement in NYHA class ( $1.3 \pm 0.4$  vs  $1.8 \pm 0.6$ ,  $p = 0.05$ ) and exercise test capacity ( $93 \pm 22$  W vs  $78 \pm 14$  W,  $p < 0.05$ ).

In spontaneous rhythm (SR), a significant inter-ventricular dyssynchrony was found ( $41 \pm 13$  ms,  $p < 0.01$ ) as well as late activation of RV lateral wall (electrosystolic delay:  $42 \pm 22$  ms vs lateral LV wall and  $49 \pm 30$  ms vs interventricular septum;  $p < 0.01$  for both). This dyssynchrony is corrected in biventricular pacing (inter-ventricular delay  $8.6 \pm 6.4$  ms electrosystolic delays respectively  $25.5 \pm 13$  ms and  $12 \pm 9$  ms,  $p < 0.01$  vs SR). RV pacing is responsible for late activation of LV lateral wall ( $36.5 \pm 30$  ms).

**Conclusion:** BVP pacing in selected patients with repaired TOF, BVP significantly improves dyssynchrony parameters. This is associated with significant improvement of clinical status.

#### P-158

##### **Follow-up after Fontan Conversion**

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**Objective:** Review of patients with Fontan conversion to assess a strategy for these patients.

**Methods:** From 1994 to 2010 twenty-eight patients (age 10.5 to 49.5 years, mean 24.2 years) with Fontan modification underwent a conversion from atriopulmonary to total cavopulmonary connection, indications for the operation were "failing Fontan" and refractory arrhythmias. The follow-up data were retrospectively extracted from the outpatient files.

**Results:** Follow-up was between 6 month and 16.5 years. The first 15 patients were operated by means of an intracardiac tunnel, after 2001 by means of an extracardiac conduit.

Total mortality was 8/28, with five early postoperative deaths and three deaths in the later follow-up after 14 months, 33 months, and 10,5 years with protein losing enteropathy in two cases and sepsis fulminans in one as cause of death. One patient had a heart transplantation 9 years after Fontan conversion.

At follow-up, there was a recurrence of arrhythmia in 15/23 patients, 10/23 patients were dependent on a pacemaker, 9 patients had atrial tachycardia of whom two required electrical cardioversion, all of them are on anti-arrhythmic medication. Maze operation was performed in four patients at the time of conversion with reoccurring of arrhythmias in three patients. Exercise tests in the first 12 months after surgery showed worse results than before surgery but improved at follow-up. At their last outpatient visit, 21/23 patients had a actual job as sign of good social re-integration.

**Conclusion:** Fontan conversion is a high risk operation in terms of perioperative and intermediate mortality. Preoperative evaluation and carefully planned anti-arrhythmic surgery is necessary to improve clinical outcome.

#### P-159

##### **Univentricular heart and body growth: are we timing the Fontan stages properly?**

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**Objective:** The staged Fontan procedure for univentricular heart palliation is designed to gradually reduce volume overload and cyanosis during infancy and early childhood. The optimal timing of this staging and its impact on somatic growth is still a matter of debate. We explored the trends in body growth related to subsequent surgical and interventional procedures and the need for heart failure treatment.

**Methods:** We reviewed 64 consecutive patients that ultimately underwent a total cavopulmonary connection (TCPC) in our centre since 1992. Serial anthropometric parameters (weight, height) were recorded from birth to latest follow-up (mean

patient FU 7,  $8 \pm 5$  years) and at each intervention (neonatal surgery, bidirectional cavopulmonary anastomosis (BCPA), TCPC, catheter treatment), and converted to z-scores. The influence of oxygen saturation, heart failure treatment and interval between surgeries on body growth were determined.

**Results:** Median age at BCPA and TCPC was 0.9 (0.4–6.4) and 3.2 (2.2–18.3) years, 8 patients underwent unstaged TCPC after previous neonatal surgery. Median z-scores for weight changed significantly after each surgical stage ( $-0.4$  at birth,  $-0.8$  at neonatal surgery,  $-2.2$  at BCPA,  $-1.1$  at TCPC, and  $-0.6$  at latest FU;  $p < 0.05$  for change between surgeries), with the largest decline awaiting BCPA, and the most marked improvement before and moderate increase after TCPC. Z-scores for height showed the same pattern up to the TCPC stage (median 0 at birth,  $-0.4$  at neonatal surgery,  $-1.5$  before BCPA,  $-0.6$  at TCPC;  $p < 0.05$  for each interval), but did not any more improve after TCPC (median  $-0.5$  at latest FU). Somatic growth at latest FU was negatively influenced by the use of heart failure treatment ( $p < 0.05$ ), but not by age at TCPC, ventricular morphology or cyanosis.

**Conclusion:** Body growth is decreased in all patients with univentricular heart palliated with a Fontan circulation. The severe growth impairment occurring before BCPA would suggest advancing this stage where possible, allowing an earlier start of catch-up growth before TCPC. Insidious heart failure after Fontan completion should be treated promptly, in view of its negative impact on somatic growth.

#### P-160

##### **Magnetic resonance imaging and cardiopulmonary test in adults after repair of tetralogy of Fallot: effects of pulmonary valve replacement**

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**Background:** Pulmonary regurgitation (PR) is a common complication after total correction for Tetralogy of Fallot. Chronic PR leads to progressive right ventricular (RV) dilatation and dysfunction, reduction of exercise capacity and increased incidence of arrhythmias and sudden cardiac death. The optimal timing for elective pulmonary valve replacement (PVR) is unknown.

In this study, MRI was used to assess the effects of PVR on RV function and PR.

Furthermore exercise capacity was evaluated by VO<sub>2</sub> peak.

**Methods and Results:** We included twenty patients (60% male, age  $29 \pm 13$  years). All patients underwent to exercise test at a median of  $8.2 \pm 4.3$  months before and  $11.4 \pm 4.1$  after PVR, with measurement of peak exercise oxygen uptake (VO<sub>2</sub> peak). Cardiac MRI was performed at a median of  $9.3 \pm 3$  months before and  $10.6 \pm 4.2$  months after PVR.

After PVR only 2 patients showed mild residual PR.

In the intervention group VO<sub>2</sub> peak didn't increased significantly (from  $27.2 \pm 7.3$  to  $28.7 \pm 6.6$  ml/min;  $p = 0.47$ ).

RV end – diastolic volume (RV – EDV) decreased from  $160.9 \pm 30.1$  mL/m<sup>2</sup> to  $90.16 \pm 11.6$  mL/m<sup>2</sup> ( $P < 0.005$ ); and RV end-systolic volume (RV – ESV) decreased from  $92.2 \pm 12.1$  mL/m<sup>2</sup> to  $42 \pm 13.8$  mL/m<sup>2</sup> ( $P < 0.05$ ).

No significant increased of RV – EF was found (from  $44.1 \pm 4.3$  to  $48.1 \pm 4.9\%$ ,  $P 0.1$ ).

**Conclusion:** In our series of adult patients with tetralogy of Fallot and severe PR undergone to PVR, our results evidence a

dramatic decreased of RV-EDV in absence of significant increased of exercise capacity, despite the timing of surgery was consistent to international guidelines.

We speculate that this findings suggest a more restrictive indications for timing for surgery of PVR.

#### P-161

##### **Activity in Adolescents with Congenital Heart Disease (CHD)**

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**Introduction:** Ability to exercise is an important contributor to and indicator of physical health. Many patients with CHD are adolescents and their attitude to exercise is variable. Exercise capacity is an important prognostic factor in acquired heart disease and may have a similar role in CHD. We aim to examine exercise ability in adolescents with major CHD compared to those with a minor diagnosis.

**Methods:** Patients aged 12–20 years were identified using the Northern Ireland regional database (HeartSuite). Participants were categorised as having major or minor CHD and divided into four diagnostic subgroups. They completed a validated questionnaire rating their exercise capacity. Participants also underwent a formal exercise stress test using the Bruce Protocol and measurement of free-living activity using an ActiGraph accelerometer. Results were analysed using parametric methods.

**Results:** 143 patients (mean age 15.6 years) consented to participate, 86 were male (60%) and 105 had major CHD (73%). Diagnostic subgroups included 39 acyanotic (27.3%), 61 acyanotic corrected (42.7%), 30 cyanotic corrected (21.0%) and cyanotic palliated 13 (9%). 134 participants (93.7%) took part in regular exercise each week, for 68 individuals (47.6%) this was more than 3 times per week. There was no significant difference in activity score between study groups. 142 participants attempted an exercise test; more complex patients performed worse at peak exercise. Exercise time for acyanotic group 11.73 mins (sd 3.74) compared to 8.26 mins (sd 4.08) in cyanotic palliated group, ( $p$  value 0.002 (1.32, 5.61)). Patients with major CHD, especially acyanotic corrected group, had significantly higher activity counts. This difference was reflected across subgroup analysis ( $p$  value 0.007) with acyanotic patients having the lowest activity scores.

**Conclusions:** The majority of young people with CHD, in this group, take part in regular exercise. Surprisingly complex patients rate themselves to be as active as those with minor CHD. While accelerometer data indicate that group may be more active day to day, they are limited in terms of peak exercise duration. Interventions targeted towards maintaining or increasing exercise capacity may confer prognostic benefits for these patients.

#### P-162

##### **Quality of life in families with a child after staged treatment for hypoplastic left heart syndrome (HLHS)**

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**Background:** In spite of increasing number of survivors with HLHS after staged Norwood operation – data about quality of life in this group of patients and their families is limited.

**Aim:** The aim of the study was to assess the quality of life of children with HLHS and their families.

**Methods:** Mothers of 52 children with HLHS completed the questionnaire to assess subjectively quality of their children's life and impact of child's illness on the family. All children (age 3–15 years) were operated at our institution.

**Results:** Problems in physical activity in HLHS patients were reported in 69%, emotional problems in 25%, educational in 12%, problems in psychomotoric development were reported in 31%. 89% of analyzed patients attend to normal schools or kindergartens, and in 100% they are accepted by peers and teachers. Good tolerance of frequent hospitalizations was reported in 75% of estimated patients.

Child illness is connected with strong parental stress (73%), and negative emotions like sadness (41%), fear and helplessness (42%). Own family support, support groups of families with the same problem and religious faith were considered as most helpful. 94% responders assessed familial atmosphere as good, in 81% child's illness strengthened parental marriage, good relations among siblings and HLHS patients were reported in 77%. Only in 29% the need of limitation in family activity was reported. Impact of child's illness on family economic situation was assessed as significantly negative in 79%.

**Conclusion:** Patients with HLHS are active members of the society, they attend to normal schools and kindergartens although their physical activity is limited. The family functioning is good although child's illness is a reason of strong parental stress and indicates familial economic problems.

Increasing number of HLHS survivors indicates the need for continuation studies concerning neurodevelopmental outcome, quality of life and family functioning in this group of patients.

#### P-163

##### Do patients benefit from Ross procedure even in long-term?

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**Introduction:** Ross procedure (ROSS) is a type of specialized aortic valve operation, where diseased aortic valve is replaced with the person's own pulmonary valve and a homograft placed in pulmonary position.

**Patients:** Between years 1997–2010, 48 patients (31M/17F, mean age  $12.9 \pm 5.3$  years) with aortic valve malformation underwent ROSS at our institution: in 29 (60.4%) patients surgery was performed due to predominantly aortic regurgitation (AR) and in 19 (39.6%) due to predominantly aortic stenosis (AS). 16 (33.3%) patients had a history of previous aortic valve (catheter or surgical) intervention ( $6.8 \pm 4.2$  years prior ROSS). **Methods:** In prospective follow-up (FU) repeated clinical evaluation and echocardiographic measurements were performed. Proximal aorta diameters (annulus, sinuses, ascending aorta) and aortic valve function; as well as pulmonary conduit function were evaluated. Need of reintervention, aortic dilatation and regurgitation were analyzed; differences between AR and AS groups were statistically evaluated.

**Results:** Short-term hemodynamic effect: in AR group left ventricle dilatation decreased significantly ( $p < 0.0001$ ) by 3 months after procedure; on the contrary in AS group left ventricle hypertrophy remained unchanged ( $p = 0.52$ ).

Long-term evaluation: at mean  $8 \pm 3.6$  years of FU, freedom of reintervention was 87.5%. Reintervention was needed in 6 (12.5%) patients (in 4 because of pulmonary homograft stenosis/in 2 because of aortic regurgitation), mean  $6.9 \pm 3.4$  years after ROSS.

In patient with FU  $> 5$  years a higher incidence of aortic root dilatation ( $p = 0.017$ ) and pulmonary homograft stenosis ( $p < 0.0003$ ) was found. Severe pulmonary regurgitation was more often present in patients who underwent ROSS at age  $> 10$  years ( $p = 0.0058$ ). In AR group compared to AS – higher incidence of aortic regurgitation ( $p = 0.01$ ) and aortic annulus dilatation ( $p = 0.023$ ) was found. Systemic hypertension was a risk factor for ascending aorta dilatation ( $p = 0.04$ ). Pre-operative aorto-pulmonary disproportion ( $> 3$  mm) and pulmonary valve dimension did not effect long-term aortic dilatation or regurgitation.

**Conclusions:** Ross procedure has a good short-term benefit and low need for reintervention. On the other hand, aortic dilatation and pulmonary conduit dysfunction can be expected to progress with time and may be the main cause for further reintervention. Patients with previously regurgitant lesion tend to more severe aortic dilatation and new aortic regurgitation.

#### P-164

##### Incidence and Etiology of Secondary Surgical Interventions on the Aorta in Marfan Syndrome

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**Objectives:** Patients with Marfan syndrome (MFS) frequently require interventions on the distal aorta. Aim of the current study was to determine incidence and etiology of secondary surgical interventions in downstream aortic segments in MFS after previous aortic repair.

**Patients and Methods:** Data were prospectively collected from 86 MFS patients fulfilling Ghent criteria that underwent a total of 136 operations and were followed at this institution between 1995 and 2010.

**Results:** Mean follow-up of survivors was  $8.8 \pm 6.8$ y, mean time-to-re-operation was  $5.5 \pm 4.6$ y. Thirty-day, 6 months, 1 year and late mortality was 3.5%, 5.8%, 7.0% and 12.8%, respectively. Four out of these 11 deaths (36%) were due to aortic rupture during late follow-up.

Seventy-eight patients (91%) primarily presented with root, ascending or arch lesions, whereas 7 patients (8%) presented with thoracoabdominal lesions. Etiology at primary presentation was acute dissection in 36% [24 (77%) type A, 7 (23%) type B] and chronic dilative disease in 64%. Secondary arch replacement had to be performed in only 6% of patients in the non-dissected, but in 36% of the dissection group ( $p = 0.0005$ ).

In the non-dissection group, 11% of patients underwent surgery in downstream aortic segments [5 out of 6 patients suffered from type B dissection in the meantime], whereas in patients after acute dissection, 48% patients had to undergo surgery on the distal aorta [42% of patients with type A and 86% of those with type B dissection] ( $p = 0.0002$ ).

**Conclusion:** In a contemporary cohort of patients with MFS, a third of the patients still presents initially with acute dissection despite the wide availability of prophylactic surgery. The current data suggests that in patients with MFS, the need for repeated surgery in downstream aortic segments is not determined by the segment of the aorta that is initially involved but rather the presence or absence of acute dissection.

**P-165****The outcome of Eisenmenger patients with trisomy 21 does not differ from patients without trisomy 21**

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*Introduction:* Several patients with trisomy 21 developed the Eisenmenger syndrome (ES) because the underlying congenital heart defect was not corrected. However, little is known about their prognosis. This study aimed at (1) identifying risk factors for worse prognosis in ES patients, and (2) evaluating whether outcome of ES patients with trisomy 21 differs from ES patients without trisomy 21.

*Methods:* Data on all Eisenmenger patients in follow-up at the pediatric and adult congenital heart disease clinic of the University Hospitals Leuven were collected for retrospective analysis. Regression analysis was performed where applicable and survival rate was compared between patients with and without trisomy 21.

*Results:* One hundred thirty-four patients (mean age at latest follow-up  $33.2 \pm 13.6$  years, 41.8% male, 44.8% trisomy 21) were included in the study. Complex lesions, right heart failure, impaired renal function, lower transcutaneous saturation and lower body mass index were predictive for impaired outcome. Mean survival of the global ES group was  $44.9 \pm 2.2$  years. However, long-term survival of trisomy 21 patients was not statistically different from patients without trisomy 21 (mean survival  $44.5 \pm 2.6$  years vs  $44.5 \pm 2.9$  years respectively,  $P = 0.80$ , log rank test).

*Conclusions:* Long term survival is markedly reduced in Eisenmenger patients. Complex lesions, right heart failure, impaired renal function, lower transcutaneous saturation and lower body mass index were related towards prognosis. However, survival of trisomy 21 patients did not differ from patients without trisomy 21.

**P-166****The long term outcome of patients with interventional treatment of pulmonary valvular atresia/stenosis**

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*Objectives:* to evaluate the long-term outcome of neonates with pulmonary atresia and intact ventricular septum (PAIVS)/critical pulmonary valvular stenosis (PVS) undergone successful transcatheter treatment.

*Methods:* The study population consists of 39 patients (pts) with successful transcatheter treatment of critical pulmonary valvular obstruction, with a minimum follow-up period after the procedure of 2 years. The surgical and interventional procedures performed after the first treatment and the clinical and echocardiographic data at follow-up were analysed.

*Results:* Among the 39 pts, 12 pts were affected by PAIVS and had undergone radiofrequency perforation followed by pulmonary valvuloplasty, 27 by critical duct-dependent PVS and had been treated with pulmonary valvuloplasty. The mean age at the time of the first treatment was  $3 \pm 4$  days and the mean duration of follow-up was  $9 \pm 4$  years. Twenty-eight pts (72%) received only interventional treatment: in 8 cases the valvuloplasty was repeated and 6 underwent ASD closure. In all pts of this group,

the O<sub>2</sub> saturation at follow up was  $\geq 95\%$ ; the Doppler mean peak pulmonary valvular systolic gradient was  $17 \pm 9$  (5–35) mmHg and 2 pts presented well tolerated moderate pulmonary insufficiency. Eleven pts (28%) required additional surgical treatment/s during the follow-up period: 6 mBT, 7 RVOT reconstruction, 4 bidirectional cavopulmonary anastomosis, 1 total cavopulmonary anastomosis. The tricuspid valve Z-score at the time of the first percutaneous treatment was  $-0.97 \pm 1.1$  in the 28 pts treated only with interventional procedures and  $-0.38 \pm 1.6$  in the 11 pts requiring also surgery ( $p$ : NS). Moreover, 9/39 pts (23%), of whom 6 treated only percutaneously, with unfavourable tricuspid or pulmonary valve annulus Z-score, achieved biventricular circulation.

*Conclusions:* In our experience transcatheter treatment is an effective technique for the decompression of the right ventricle in neonates with critical pulmonary valvular obstruction. The late outcome is often unpredictable and depends on the effectiveness of the intervention as well as on the anatomic and functional adequacy of the right ventricle.

**P-167****Detrimental effects of chronic non-systemic ventricular pacing in patients with atrial switch for transposition of the great arteries**

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*Introduction:* Patients with transposition of the great arteries (TGA) treated by atrial switch are at risk of systemic ventricular dysfunction. In addition, these patients often develop conduction disorders and/or cardiac arrhythmias leading to pacing device implantation. The aim of our study was to evaluate the effects of chronic non systemic ventricular pacing in these patients compared to a control group of patients with atrial switch for TGA.

*Methods:* 11 patients with permanent or predominant (>90% of time) left ventricular pacing were compared to a control group of 31 non paced patients. Each patient's clinical status was analysed and then underwent -1- complete echocardiographic analysis with dyssynchrony measures, -2- exercise test with peak O<sub>2</sub> consumption, -3- Blood Brain Natriuretic Peptide (BNP) level measure.

*Results:* Both groups were comparable (paced vs, non paced: mean age 28.1 vs 25.3, men 63 vs 71%, weight 66.7 vs 66.3, all ns) Patients with left ventricular pacing have significantly worse clinical status (NYHA class  $1.9 \pm 0.3$  vs  $1.16 \pm 0.2$   $p < 0.01$ ) and exercise test performance (maximum performance  $100 \pm 30$  W vs  $120 \pm 32$  W, peak oxygen consumption  $22 \pm 6$  vs  $27 \pm 7$ ; both  $p < 0.05$ ). Echocardiographic parameters showed significantly reduced right ventricular systolic function parameters (shortening fraction  $27 \pm 11\%$  vs  $33 \pm 10\%$ , dP/dt  $1034 \pm 250$  cm/s vs  $891 \pm 470$  cm/s, both  $p < 0.05$ ) and increased inter and intraventricular dyssynchrony (respectively  $99 \pm 10$  ms vs  $25 \pm 9$  ms and  $70 \pm 29$  ms vs  $21 \pm 15$  ms, both  $p < 0.01$ ). BNP levels are respectively  $137 \pm 176$  and  $103 \pm 168$  (ns) in paced and non paced patients.

*Conclusion:* Permanent left ventricular pacing in patients with atrial switch for TGA is deleterious with reduced exercise capacity, worse functional status and significant dyssynchrony along with altered systemic right ventricular function. Cardiac resynchronization therapy should be considered as an alternative to conventional pacing in patients with systemic ventricular dysfunction.



**P-168****Impaired fibrinogen function in patients with cyanotic congenital heart disease**

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**Introduction:** Patients with cyanotic congenital heart disease (CCHD) have haemostatic abnormalities associated with bleeding and thrombo-embolic events. These haemostatic abnormalities are not fully understood, but recent studies indicate that elevated hematocrit and fibrinogen function may be of importance.

The aim of this study was to characterize haemostatic abnormalities in CCHD patients and examine how these are affected by increased hematocrit.

**Methods:** In a prospective study fifty adult CCHD patients had hematocrit, platelet count, and fibrinogen concentration examined. Thrombelastography (standard TEG) values R (initiation of coagulation), Angle (clot build-up), MA (maximal clot strength), as well as TEG Functional Fibrinogen (TEG FF) assay evaluating fibrinogen function (FLEV) was performed. In addition TEG FF gave information about fibrinogen and platelet contribution to clot formation and strength.

**Results:** Average hematocrit was  $56 \pm 8\%$  and platelet counts in the lower normal range. Standard TEG revealed prolonged R, reduced Angle but normal MA indicating a hypocoagulable condition with impaired clot formation but preserved clot strength. Interestingly the standard TEG R, Angle and MA values were correlated to elevated hematocrit, indicating that elevated hematocrit caused a hypocoagulable state. Despite of high levels of plasma fibrinogen in patients with elevated hematocrit, TEG FF demonstrated that FLEV was diminished particularly affecting clot formation but also strength. On the other hand the platelet function was normal despite of low platelet count.

**Conclusion:** Patients with CCHD are hypocoagulable mainly due to impaired fibrinogen function. Despite a low platelet count, platelet function seems to be normal in CCHD patients. Haemostasis, and especially fibrinogen function, is negatively affected by elevated hematocrit.

**P-169****The hematocrit – an important factor causing impaired haemostasis in patients with cyanotic congenital heart disease**

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**Introduction:** Patients with cyanotic congenital heart disease (CCHD) have haemostatic abnormalities, which result in an increased risk of bleeding. The cause is unknown, but recent studies have indicated that an elevated hematocrit, which is present in cyanotic patients, could be an important factor. This is contrary to the previous assumption that elevated hematocrit is associated with increased risk of thrombosis. The aim of this study was to characterize haemostatic abnormalities, examine how changes in hematocrit affect the haemostatic profile and

whether a hematocrit reduction could terminate bleeding in CCHD patients.

**Methods:** This was a prospective, multicenter study. The haemostatic profile was characterized in ninety-seven clinically stable CCHD patients. To evaluate the influence of hematocrit on the haemostatic profile, 21 of the patients underwent phlebotomy and 16 patients received treatment with an iron supplement. Furthermore ten patients with hemoptysis underwent phlebotomy. The haemostatic profile was reevaluated after interventions. The haemostatic profile consisted of hematocrit, platelet count, thrombelastograph (TEG), and plasma coagulation analysis.

**Results:** TEG revealed that patients with CCHD and elevated hematocrit were hypocoagulable. Furthermore a positive correlation between elevated hematocrit and hypocoagulability was present. Interventions such as phlebotomy and treatment with supplemental iron causing significant hematocrit changes confirmed the correlation between hematocrit and the haemostatic profile. Finally a hematocrit reduction by phlebotomy successfully terminated the hemoptysis in all ten CCHD patients.

**Conclusion:** Patients with CCHD and elevated hematocrit are hypocoagulable. Haemostasis seems to be affected by changes in hematocrit and phlebotomy may be a possible treatment of bleeding in CCHD.

**P-170****In operated fallot is RV end-diastolic volume >170 ml/m<sup>2</sup> a valid cut-off for indication to pulmonary valve replacement?**

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**Introduction:** Patients with Tetralogy of Fallot (TOF) repaired by means of a transannular patch, suffer of pulmonary regurgitation leading to right ventricle (RV) dilatation. Several groups have recently demonstrated that if these pts are re-operated for pulmonary valve implant (PVR) since indexed RV end-diastolic volume (RVdVol) exceeds  $170 \text{ mL/m}^2$ , the RV volumes do not decrease to normal values. However the beneficial hemodynamic effects of PVR still have to be weighted against the need for re-operation for valve failure. Moreover in TOF to aim to a “normal” RV previously “violated” by means of an infundibulotomy and other surgical procedures is a simple surrogate but probably unfit for them.

**Aim:** to compare the clinical outcome and instrumental data of two matched repaired TOF population, differing by RVdVol ( $\geq 170 \text{ mL/m}^2$  vs  $< 170 \text{ mL/m}^2$ ).

**Methods:** From our database we identified 27 TOF >15 y.o. with transannular patch as primary repair, not yet re-operated for PVR and with RVdVol  $\geq 170 \text{ mL/m}^2$ . This group of pts (group 1) has been matched for sex, age and age at repair against 32 TOF with the same characteristic above mentioned, but with RVdVol  $< 170 \text{ mL/m}^2$  (group 2). All of them were evaluated by cardiacMR, echocardiography, cardiopulmonary exercise test. Clinical adverse outcomes were also recorded.

**Results:** Only pulmonary regurgitation fraction resulted significantly different between group 1 and 2 ( $47 \pm 11\%$  vs.  $37 \pm 10\%$  respectively  $p < 0.01$ ). Conversely other parameters resulted not significantly different:  $\text{Vo}_2/\text{kg}/\text{min}$   $24 \pm 7 \text{ mL}/\text{kg}/\text{min}$  vs.

23.5 ± 6 respectively, RVEF 50.5 ± 7.2% vs. 52 ± 6.4% respectively. Eleven pts (19%), 5 from Group 1 (18%) and 6 from Group 2 (19%) experienced adverse events: 2 sustained ventricular tachycardia, 8 major atrial arrhythmias, 1 worsening in NYHA functional class. RVdVol ≥170 ml/m<sup>2</sup> was not associated to adverse event (O.R. 1.1 95% CI: 0.298 to 4.105).

**Conclusion:** From our data, the two groups didn't differ in term of adverse event and/or instrumental finding of RV dysfunction. We believe that in TOF the RVdVol and the timing of PVR should be evaluated in combinations with several other clinical and instrumental parameters.

#### P-171

##### **High percent of reinterventions in patients with aortic coarctation repair**

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**Aim:** To study late outcome after surgical repair in adult patients with simple aortic coarctation.

**Patients and Methods:** Forty-one patients (mean age: 28 ± 8 years, female: n = 12, male: n = 29) who underwent surgical repair for simple aortic coarctation and have regular follow-up data available in our centre were recruited for a retrospective analysis. Patients with complex left-sided cardiac anomaly or interrupted aortic arch were excluded.

**Results:** Major associated congenital anomaly was bicuspid aortic valve (n = 18, 44%). Mean age at first repair was 6 ± 5 years. The type of first repair was: patch repair (n = 25), end-to-end anastomosis (n = 13) and interposition graft (n = 3). At a mean age of 15 ± 11 years, 46% of patients had reintervention (surgery: n = 7, balloon dilatation: n = 11, stent implantation: n = 1). Reason for re-intervention was: re-coarctation (n = 16), aneurysm (n = 1), dissection (n = 1) or aortectasia (n = 1). One patient had aortic rupture at the time of percutaneous intervention requiring urgent operation, which was performed successfully. Twelve percent of patients required a third intervention (surgery: n = 4 and endovascular graft stent: n = 1). Bicuspid aortic valve or type of first surgery did not correlate with their need for reintervention. At the last follow-up 44% of patients were hypertensive and were on at least one antihypertensive medication (n = 18). Also a high percent of patients were under investigation because of suspicion for recoarctation, as by arm-leg systolic blood pressure difference of >20Hgmm (53%) and systolic pressure gradient of > 30 mmHg at the site of coarctation repair (32%). There were no patients with known or suspicion for coronary disease.

**Conclusions:** A high percent of young patients with history of aortic coarctation repair required reintervention, presents with hypertension and are at suspicion for recoarctation. Close follow-up in these patients is mandatory.

#### P-172

##### **Quality of life of patients after percutaneous closure of atrial septal defect**

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**Objective:** This study analyzes the quality of life (QoL) of patients after percutaneous closure of atrial septal defect (ASD) and the possible impact of age, psychological and functional status assessed by self reported questionnaires.

**Methods:** All 199 patients aged older than 16 years old, registered at the congenital heart disease center, who had undergone

percutaneous closure of ASD between May 2000 and December 2008, were sent a questionnaire package containing the SF-36 health survey (health related quality of life), the Beck Depression Inventory Scale (BDI), the Zung Self rating Depression Scale (Zung SDS) and the Duke's Activity Status index (DASI). Questionnaires were correctly completed by 89 patients (44,7%), 56 women and 33 men, median age 42,3 ± 17,8 (range 16 to 76,2) years. Patients were assigned to one of three categories based on their age during the study; group A (young adults aged 16 to 24 years), group B (adults aged 25 to 60) and group C (elderly aged older than 60 years). The scores of QoL of each group were compared within them and with age matched normative data of the greek population.

**Results:** In all SF-36 health dimensions patients showed excellent scores which are comparable to the normative data (p < 0,025). Patients with depressive symptoms as estimated by both depression scales had significant lower scores in all dimensions of QoL (p < 0,021). All SF-36 dimensions correlated significantly with functional status as assessed by DASI (p < 0.005). Age affected physical functioning (p = 0,013), bodily pain (p = 0,005) and general health perception (p = 0,018), as in normal population.

**Conclusions:** QoL in patients following percutaneous closure of ASD is excellent and comparable to standard population. As expected, older patients had poorer levels of physical functioning and overall general health perception compared to their younger counterparts, similar though to their normal peers. Depressive symptoms and impaired functional capacity was also associated with poorer QoL. Our data demonstrate that ASD closure, even at advanced age, does not have a negative effect in any of the QoL domains.

#### P-173

##### **Changes of myocardial deformation and dyssynchrony in children with hypoplastic left heart syndrome before and after the Norwood operation assessed by 2D speckle tracking**

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**Background:** 2D speckle tracking (2DST) is an echocardiographic technique that is independent of ventricular geometry and the angle of insonation but dependent on loading conditions of the heart. The purpose of our study was to investigate differences in deformation parameters and intraventricular dyssynchrony (ID) before and at steady state after the Norwood operation (NO) in neonates with hypoplastic left heart syndrome (HLHS) using 2DST.

**Methods:** On echocardiograms before and 23 ± 15 days after the NO, we compared global and regional peak systolic longitudinal strain (S) and strain rate (SR) as well as ID of 33 HLHS patients. ID was calculated as the standard deviation of the time intervals from the beginning of the QRS interval to peak S for the 6 segments of the right ventricle (Yu index) as well as the delay of time to peak S between basal septal and lateral walls.

**Results:** Global S (-18.3 ± 3.6 vs. -16.8 ± 3.8%, p = 0.02) and global SR (-1.6 ± 0.3 to -1.2 ± 0.3 s<sup>-1</sup>, p < 0.0001) decreased significantly. Regional S decreased significantly in the apical (-24.1 ± 4.8 vs. -19.6 ± 6.4%, p = 0.003) and mid lateral segments (-21.8 ± 4.2 vs. -19.2 ± 3.9%, p = 0.01) while

regional SR decreased significantly in all but the basal septal segments. ID remained unchanged (Yu index  $38 \pm 15$  vs.  $41 \pm 20$  ms, wall-to-wall delay  $64 \pm 46$  vs.  $60 \pm 51$  ms). Arterial oxygen saturation and heart rate were lower after NO.

**Conclusion:** 2DST parameters of global and regional RV deformation were lower after the NO while ID remained similar. Factors influencing this decrease most likely include changes in loading conditions, hemodynamic parameters and medication.

#### P-174

##### Comparison of myocardial deformation and dyssynchrony in children with left and right ventricular morphology after the Fontan operation assessed by 2D speckle tracking

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**Background:** Right ventricular morphology has been found to be associated with poorer clinical outcome and ventricular function in single ventricle patients after Fontan palliation. Using 2-dimensional speckle tracking (2DST), a method that can assess ventricular function independent of ventricular geometry, the aim of our study was therefore to compare global and regional deformation and intraventricular dyssynchrony (ID) between children with left and right ventricular morphology after Fontan operation.

**Methods:** We compared global and regional longitudinal strain (S) and strain rate (SR) as well as ID between 29 patients with systemic right (group 1: age  $7.7 \pm 2.7$  y,  $\Delta t$  Fontan to echo  $5.3 \pm 3.0$  y) and 22 patients with systemic left (group 2: age  $7.8 \pm 4.8$  y,  $\Delta t$  Fontan to echo  $4.6 \pm 4.2$  y) ventricles. ID was calculated as the standard deviation of the time intervals from the beginning of the QRS interval to peak S for the 6 segments of the right ventricle (Yu index) as well as the delay of time to peak S between basal septal and lateral walls.

**Results:** Global S ( $-18.5 \pm 3.5$  vs.  $-17.9 \pm 3.2\%$ ,  $p = \text{NS}$ ) and global SR ( $-1.0 \pm 0.2$  vs.  $-1.0 \pm 0.2$  s<sup>-1</sup>,  $p = \text{NS}$ ) did not differ between groups. Regional S ( $-8.7 \pm 8.6$  vs.  $-14.7 \pm 6.7\%$ ,  $p = 0.008$ ) and SR ( $-0.7 \pm 0.4$  vs.  $-1.0 \pm 0.3\%$ ,  $p = 0.002$ ) in the basal septal segment were lower in group 1 while regional S was higher in group 1 in the apical septal segment ( $-23.5 \pm 8.0$  vs.  $-18.4 \pm 5.9\%$ ,  $p = 0.02$ ). ID was not different (Yu index  $43 \pm 19$  vs.  $40 \pm 24$  ms, wall-to-wall delay  $58 \pm 49$  vs.  $54 \pm 47$  ms).

**Conclusion:** Despite minor regional differences, overall ventricular deformation and dyssynchrony was not different between morphologic right and left ventricles. These findings may reflect similar adaptation of both ventricular morphologies to the single ventricle circulation in our cohort, albeit relatively early after Fontan surgery.

#### P-175

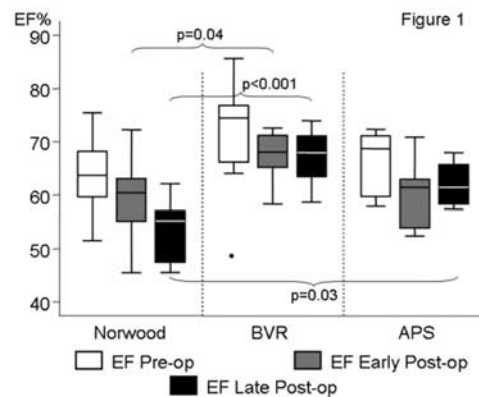
##### Progressive Contractile Dysfunction After Norwood Procedure Compared to Biventricular Repair and Isolated Aortopulmonary Shunt

Gentles T.L. (1), Cowan B. (2), Occlshaw C. (1), Beca J. (1), Finucane A.K. (1), Young A.A. (2)  
Starship Children's Hospital, Auckland, New Zealand (1); University of Auckland, Auckland, New Zealand (2)

**Introduction:** Ventricular dysfunction is a common cause of morbidity and mortality after the Norwood operation, but the mechanism is ill defined.

**Methods:** We used cardiac MRI to contrast changes in mechanical characteristics of the Norwood (NOR) ventricle in 9 patients before, one week (early) and 2 months after operation (late) with age-matched patients undergoing biventricular repair (BVR) ( $n = 8$ ) and aortopulmonary shunt (APS) ( $n = 7$ ). Systemic ventricle was RV in 8/9 NOR, 0/8 BVR and 1/7 APS.

**Results:** Systemic ventricle ejection fraction (EF) in NOR was similar to BVR and APS before operation but deteriorated progressively from before to late ( $p = 0.003$ ), and was lower than both groups at late follow-up (Figure 1). Before operation end-systolic radius/thickness ( $r/T$ ), an index of afterload, was elevated in NOR compared to BVR and was similar to APS (Table). This pattern was unchanged early and late after operation. Blood pressure measurements were not different between groups.



	Before			Early			Late		
	NOR	BVR	APS	NOR	BVR	APS	NOR	BVR	APS
$r/T$	1.9*	1.4	1.7	1.7*	1.2	1.7*	1.9*	1.3	1.6

\* $p < 0.05$  vs. BVR

**Conclusion:** In the NOR group, progressive deterioration in ventricular function occurs in the context of elevated, but stable afterload. This observation, together with the absence of dysfunction in the APS group despite a similar magnitude of afterload excess, is indicative of myocardial damage and may have important implications for long-term prognosis.

#### P-176

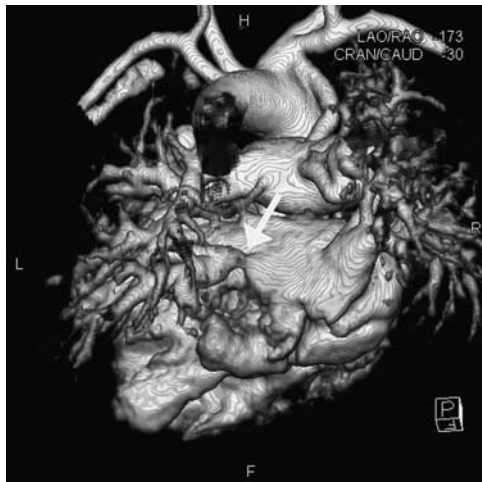
##### Assessment of Pulmonary Veins after Atrio-Pericardial Anastomosis by Magnetic Resonance Imaging

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**Objectives:** The atrio-pericardial anastomosis (APA) or "sutureless repair" uses a pericardial pouch to create a large communication between the left atrium and the pulmonary venous tributaries, avoiding direct suturing of the pulmonary veins. It is hoped that this technique leads to a reduction in the occurrence of postoperative stenosis. Post-operative imaging is routinely performed by echocardiography but cardiac magnetic resonance (CMR) offers excellent anatomical imaging and quantitative information about pulmonary blood flow. We sought to

determine the diagnostic value of CMR for assessing pulmonary vein anatomy after APA.

**Methods:** This retrospective study evaluated all consecutive patients between October 1998 and January 2010 after either a primary or secondary APA followed by a post-repair CMR. The imaging protocol included angiography and phase contrast flow velocity mapping of the pulmonary arteries and veins. The CMR findings were compared to those on echocardiography.



**Results:** Of 103 patients who had an APA at our institution, 31 patients had an analyzable CMR study. Out of these patients PV stenosis was suspected in 17 patients (55%) prior to CMR, by either clinical examination, chest radiography, echocardiography or a combination of the above. Echocardiography detected pulmonary vein stenosis in 13 out of the 17 (76%). The average time to CMR was  $24.6 \pm 32.5$  months post-repair. Echocardiographic findings were confirmed by CMR in 12 patients. There was incomplete imaging by echocardiography in 7 patients and underestimation of pulmonary vein restenosis in 12, when compared to CMR. In total, 19/31 patients (61%) from our cohort had significant stenosis following APA as assessed by CMR (the figure shows left lower pulmonary vein stenosis by contrast-enhanced angiography). Our data suggest that at least 18% (19/103) of all patients had significant obstruction post-repair.

**Conclusions:** Echocardiography incompletely images or underestimates the severity of obstruction in a significant proportion of patients following APA, as compared with CMR. Pulmonary vein stenosis remains a sizable complication after repair, even using APA.

#### P-177

##### Cardiac function after repair of tetralogy of Fallot: how are the atria performing?

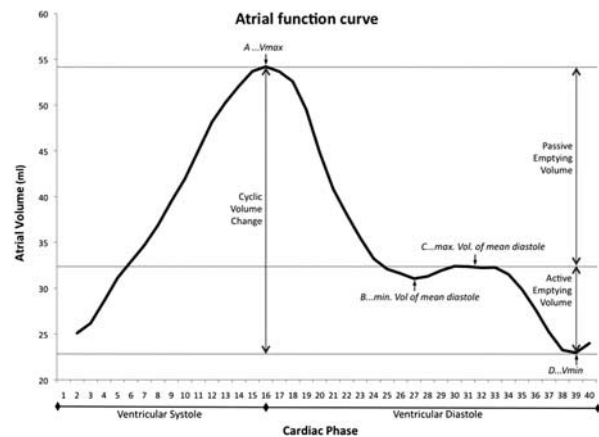
Wöhlmuth C., Vöser E., Kellenberger C.J.,

Valsangiacomo Buechel E.R.

University Children's Hospital Zurich, Switzerland

**Introduction:** During the cardiac cycle the atria function as reservoir, conduit and active pump and are critical for ventricular filling. Knowledge about atrial performance in children and in congenital heart disease is scarce.

We sought to evaluate right and left atrial size and function in patients with repaired tetralogy of Fallot (TOF).



**Methods:** Cardiovascular magnetic resonance (CMR) was performed prospectively in 12 patients after TOF repair and with significant pulmonary regurgitation. The mean age was  $16.7 \pm 6.1$  yrs, weight  $50.9 \pm 14.9$  kg. The control group consisted of 10 healthy volunteers, age  $18.8 \pm 6.8$  yrs, weight  $52.3 \pm 20.8$  kg. Steady state free precession images were acquired in a short axis plane covering both atria and both ventricles. The atrial passive and active emptying volumes, atrial emptying fraction, cyclic volume change, total atrial filling fraction and conduit volume were calculated from the volume/time curves obtained (figure). Phase contrast cine images were acquired perpendicularly to the inflow of both AV-valves. Blood flow profile across the AV-valves (E/A ratio) was used to depict subjects with ventricular diastolic dysfunction. Data were compared between patients and controls, as well as between patients with normal and abnormal ventricular diastolic function.

**Results:** In patients after TOF repair the right atrium showed an increased minimal volume at end-diastole ( $p < 0.01$ ) and increased minimal and maximal volumes during mid-diastole ( $p < 0.05$ ). Cyclic volume change ( $p < 0.05$ ), total atrial filling fraction ( $p < 0.01$ ) and passive emptying volume and fraction ( $p < 0.05$ ) were significantly decreased compared to controls. No significant difference was found for active emptying volume and fraction. In the left atrium the passive emptying fraction was the single decreased parameter ( $p < 0.05$ ) in the patient group.

Patients with a reversed E/A ratio across the tricuspid valve, representing diastolic dysfunction of the right ventricle, presented an increased conduit volume ( $p < 0.05$ ). Cyclic volume change, total atrial filling fraction, passive emptying volume and passive emptying fraction were slightly decreased, without reaching statistical significance.

**Conclusions:** Patients after TOF repair and with significant pulmonary regurgitation present an impaired right atrial function compared to controls. Right ventricular diastolic dysfunction causes even more distinctive changes of atrial parameters.

#### P-178

##### Sildenafil for successful recruitment of lost pulmonary capillaries after Fontan surgery

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**Introduction:** The status of pulmonary circulation is essential for outcome after Fontan procedure. Elevated pulmonary pressure,



vascular resistance, small size of pulmonary arteries and significant artery branch stenosis reflect some of the main problems.

**Case report:** A 5 year 8 months old boy (17,1 kg, 112 cm) with complex single ventricle morphology and coactation underwent fenestrated Fontan completion and stent implantation because of left pulmonary branch stenosis. 1 year later angiography and MRI revealed nearly no blood flow to the left lung with functional loss of capillary vascular structures. 4 months after initiation of oral Sildenafil therapy repeated MRI and angiography proved better hemodynamics, recovered lung perfusion and recruitment of the capillary vascular bed. There was significant clinical benefit.

**Conclusion:** Our case emphasizes that in case of functional loss of capillary vascular bed in non-pulsatile pulmonary perfusion recruitment of arteriolo-capillary bed is possible by oral treatment with PDE 5 inhibitor. Sildenafil was well tolerated without any adverse effects and caused significant hemodynamic and clinical improvement. It seems to be a safe and serious therapeutic option in case of functional loss of arteriolo-capillary bed.



Left: SVC-angiography with patent LPA stents but no flow to the left lung. Right: Recovered arteriolo-capillary bed after sildenafil treatment.

#### P-179

##### Bi-ventricular Diastolic Function by Tissue Doppler Imaging Correlates with Severity of Pulmonary Arterial Hypertension in Children

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**Introduction:** Pulmonary arterial hypertension (PAH) is a progressive disease resulting in a rise in pulmonary vascular resistance (PVR) and pulmonary artery pressure (PAP), and subsequently in right ventricular failure. While evidence of left ventricular diastolic dysfunction has been linked with PAH in adults, this association remains unclear in the pediatric population. The purpose of this study was to determine the association of tissue Doppler imaging (TDI) measures of bi-ventricular diastolic dysfunction when compared to invasive measurements of PAH in pediatric patients.

**Methods:** 43 children with PAH (median age 11.6 yr, range 1 mo – 21 yr) had echocardiograms and cardiac catheterizations within 7 days. Measures of myocardial longitudinal velocities by TDI included early diastole (E') and the ratio of early to late diastole (E'/A') at the lateral tricuspid, septal, and mitral valve annuli from 4-chamber view. Catheterization data included: mean PAP, PVR, pulmonary capillary wedge pressure (PCWP), right atrial pressure (RAP), and cardiac output. TDI measures were correlated against all hemodynamic measures and then evaluated for their predictive value using Receiver Operating Characteristic (ROC) analysis of E'/A' to identify patients with  $\geq$  moderate PAH as defined by mean PAP  $\geq$  35 mmHg.

**Results:** PAP and PVR, but not RAP, PCWP, or cardiac output, correlated with E'/A' ratio at the lateral tricuspid, septal, and mitral valve annuli (see Table). There was no significant correlation of the E' velocity alone. Filling pressures at catheterization were not significantly elevated (RAP 2–10 mmHg, PCWP 1–14 mmHg). ROC analysis demonstrated that tricuspid E'/A' was the best predictor of moderate PAH (AUC 0.75).

	Tricuspid E'/A'	Septal E'/A'	Mitral E'/A'
mean PAP	p < 0.001, r = 0.52	p < 0.01, r = 0.4	p < 0.001, r = 0.47
PVR	p < 0.001, r = 0.62	p < 0.01, r = 0.42	p < 0.001, r = 0.49
RAP	p < 0.14, r = 0.22	p < 0.11, r = 0.25	p < 0.08, r = 0.27
PCWP	p < 0.34, r = 0.15	p < 0.59, r = 0.08	p < 0.63, r = 0.07
Cardiac Output	p < 0.44, r = 0.16	p < 0.83, r = 0.04	p < 0.13, r = 0.29

**Conclusions:** TDI-derived E'/A' ratio correlated with direct catheterization measurement of PAH compared to E' velocity alone, suggesting the presence of compensatory atrial contraction in PAH. Bi-ventricular diastolic dysfunction significantly correlated with severity of PAH, suggesting that TDI measures may be an early sensitive marker for subclinical changes in PAH disease severity. Tricuspid E'/A' was a strong predictor of moderate or greater than moderate PAH.

#### P-180

##### Prediction of pressure gradient in aortic coarctation by computational fluid-dynamic simulations

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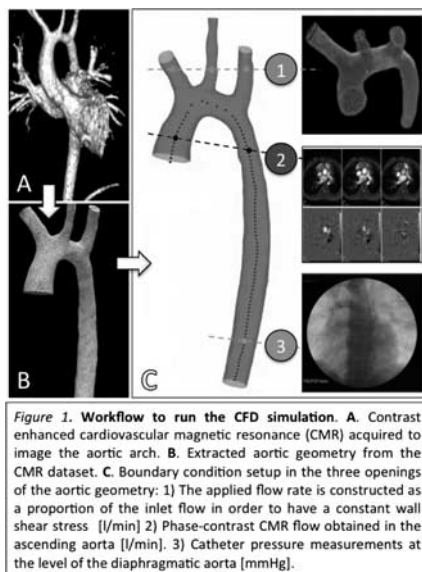
Imaging Sciences. King's College London, London, United Kingdom (1); St. Thomas & Guy's Hospitals. Evelina Children Hospital. London. United Kingdom (2); Department of Cardiovascular Science, Medical Physics Group, University of Sheffield, Sheffield, United Kingdom (3); Leiden University Medical Centre, Leiden, Netherlands (4)

**Introduction:** Current clinical assessment of borderline aortic coarctation may involve invasive catheterization pressure measurements to evaluate the pressure gradient at rest and during isoprenaline pharmacological stress. The purpose of this study is to predict the aortic pressure distribution in patients with aortic coarctation at rest and pharmacological stress using a transient rigid-walled computation fluid dynamics model (RW-CFD).

**Methods:** The study cohort comprises 7 patients (age  $19 \pm 3$  years, mean  $\pm$  standard deviation) with native or recurrent aortic coarctation and 3 control patients (age  $3 \pm 2$  years) with healthy aortic arches, who underwent both CMR and catheterization at rest and pharmacological stress. The model workflow (Figure 1) requires, as input parameters, the aortic geometry, extracted from a CMR 3D gadolinium contrast-enhanced sequence, and definition of the three boundary conditions (1) Ascending aortic root: The inlet flow is extracted from the phase-contrast CMR flow. (2) Supra-aortic vessels: The flow rate is calculated as a proportion of the inlet flow based on the assumption of a constant wall shear stress (3) Diaphragmatic aorta: The pressure waveform is extracted from the invasive catheter data. The clinical invasive aortic pressure gradients were compared with the

predicted pressure distribution along the centreline in the RW-CFD model at the time of peak flow (Table 1).

Study number	Heart rate [bpm]	$\Delta P$ Clinical [mmHg]	$\Delta P$ CFD [mmHg]	Difference in $\Delta P$ (CFD - Invasive) [mmHg]
<b>REST CONDITION</b>				
AoCo-1	48	23 $\pm$ 3	22	-1 $\pm$ 3
AoCo-2	86	18 $\pm$ 3	5	-13 $\pm$ 3
AoCo-3	69	12 $\pm$ 4	9	-2 $\pm$ 4
AoCo-4	81	10 $\pm$ 2	8	-2 $\pm$ 2
AoCo-5	60	20 $\pm$ 2	31	11 $\pm$ 2
AoCo-6	47	9 $\pm$ 2	6	-3 $\pm$ 2
AoCo-7	51	7 $\pm$ 3	8	1 $\pm$ 3
<b>STRESS CONDITION</b>				
AoCo-1	150	39 $\pm$ 6	54	18 $\pm$ 6
AoCo-2	136	40 $\pm$ 10	23	-17 $\pm$ 10
AoCo-3	130	64 $\pm$ 6	42	-22 $\pm$ 6
AoCo-4	140	66 $\pm$ 4	44	-22 $\pm$ 4
AoCo-5	102	52 $\pm$ 7	78	26 $\pm$ 7
AoCo-6	141	37 $\pm$ 7	58	21 $\pm$ 7
AoCo-7	119	69 $\pm$ 7	179	110



**Results:** For patients with aortic coarctation, during pharmacological stress, there was an increase in both heart rate (68  $\pm$  22 bpm) and invasive pressure gradient drop across the coarctation (38  $\pm$  18 mmHg, Table). The RW-CFD model predicted accurately the pressure drop at rest (-1  $\pm$  7 mmHg), and gave a moderate agreement at stress 16  $\pm$  46 mmHg (Table).

**Conclusion:** For patients with aortic coarctation, the RW-CFD simulations accurately predict the pressure gradient at rest and give indication of the gradient severity during stress, with no predicted gradient in control patients. These preliminary results are promising and represent the first step towards an image-based fluid-solid-interaction CFD analysis. In the future, it is envisaged that CFD models could be based on a patient-specific, non-invasive and non-ionising radiation assessment such as CMR in order to predict the hemodynamic conditions in the aorta and avoid invasive cardiac catheterization.

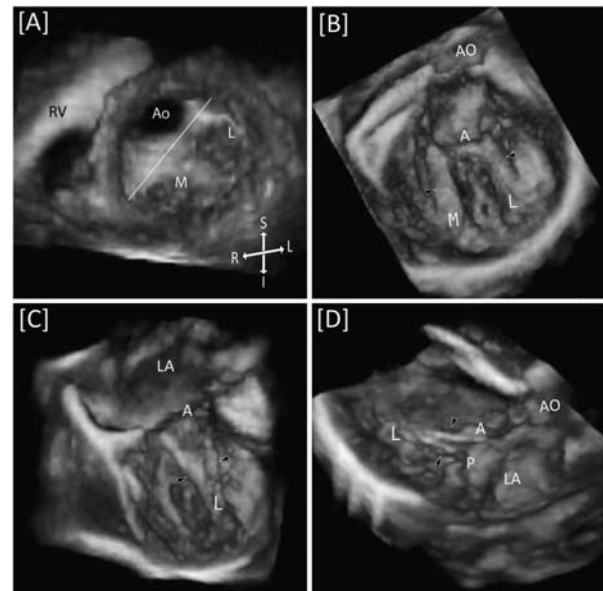
#### P-181

##### Three-dimensional echocardiographic features of parachute mitral valve

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**Introduction:** Precise delineation of the anatomy is essential to successful mitral valve (MV) repair. In recent years, three-dimensional (3D) echocardiography has emerged as an important technique for evaluation of abnormalities of the atrioventricular valves. Unique views of subvalvar MV apparatus can be obtained by 3D-echocardiography which are not achievable by standard cross-sectional two-dimensional (2D) echocardiography.



**Case report:** We present a 3D echocardiography comparison between a normal and parachute MV for teaching purposes to demonstrate the anatomy of the papillary muscle and chordal support apparatus.

**Results:** Panel A shows a short axis projection of a normal MV. By cropping a plane (line) anterior to the medial (M) and lateral (L) papillary muscles, a novel 3D projection is achieved in Panel B, showing the anterior MV leaflet (A), chordal attachments (arrows) and papillary muscles.

Analogous projection in a parachute MV (Panel C) shows both the medial and lateral margins of the anterior MV leaflet (A) with chordal attachment (arrows) to a single lateral papillary muscle (L). Panel D illustrates a long axis view of the left ventricle, revealing the attachments of the lateral aspect of the anterior and posterior MV leaflets to the single lateral papillary muscle (Please, see supplemental videos 1-3).

**Conclusions:** Identification of a parachute MV is important for surgical planning. Conventional echocardiography infers the presence of a parachute MV by the presence of a single papillary muscle but cannot demonstrate all valvar attachments in a single plane. The complementary projections which are now achievable using 3D-echocardiography facilitate, in our opinion, an improved understanding of the anatomy of chordal attachment in this lesion.

#### P-182

##### Evaluation of cardiac functions by tissue Doppler echocardiography in children with familial Mediterranean fever

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**Objectives:** Although familial Mediterranean fever (FMF) may carry a potential for cardiovascular disorders because of sustained inflammation during its course, there have been limited numbers of studies investigating the cardiac functions in pediatric patients. The aim of this study was to assess the both ventricular diastolic function using conventional echocardiography and tissue Doppler imaging in children with FMF.

**Methods:** The study population included 25 patients with FMF (12 females; mean age:  $11.8 \pm 5.30$  years) and 23 healthy subjects as controls (13 females; mean age:  $9.88 \pm 3.69$  years). All the patients enrolled in the study fulfilled the clinical criteria for FMF. All the patients were homozygous for mutations in FMF gene. The both ventricular functions were measured using echocardiography comprising standard M-mode and conventional Doppler as well as tissue Doppler imaging during an attack-free period.

**Results:** C-reactive protein and fibrinogen levels were significantly higher in FMF patients as compared to healthy controls ( $p = 0.01$ ). The conventional echocardiographic parameters with myocardial performance index were in normal ranges and similar in FMF patients and controls ( $p > 0.05$ ). However, right ventricular diastolic dysfunction was observed in FMF patients documented by tissue Doppler imaging ( $p = 0.03$  for E't and A't wave ratio).

**Conclusions:** We have demonstrated that although left ventricular functions were comparable in the patients and the healthy subjects, right ventricular diastolic function indices were impaired in FMF patients by using tissue Doppler imaging during childhood. Impaired right ventricular diastolic function may be an early manifestation of cardiac involvement in pediatric patients with FMF.

#### P-183

##### **Measurement of Coronary Flow Velocity and Coronary Flow Reserve During Stress Echocardiography in Pediatric Heart Transplant Patients**

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Children's Heart Centre, British Columbia Children's Hospital and The University of British Columbia, Vancouver, Canada

**Introduction:** Coronary flow velocity (CFV) and coronary flow reserve (CFR) by transthoracic Doppler echocardiography represents a novel non-invasive tool for the assessment of coronary allograft vasculopathy (CAV) in adult heart transplant patients. However, no data currently exists describing the pattern of CFV or CFR in pediatric heart transplant patients (PHTP) using non-invasive imaging during stress. In this pilot study, we sought to determine the feasibility of using this technique to measure CFV and CFR during stress testing in this group of patients.

**Methods:** Measurements of resting and peak CFV and CFR (peak CFV/resting CFV) were obtained from transthoracic Doppler echocardiograms performed during semi-supine cycle exercise (SSCE) ( $n = 8$ ) or dobutamine stress (DSE) ( $n = 10$ ) testing in a total of 14 PHTP. In 3 of these patients, 2 coronary arteries were examined during a single test. The findings were correlated with each patient's rejection status obtained from their myocardial biopsy histopathology reports and coronary artery status imaged by angiography. A normal CFR value was defined as  $>2$  times the resting value.

**Results:** CFV and CFR were measured in the LAD ( $n = 4$ ), left main ( $n = 11$ ), RCA ( $n = 3$ ) and circumflex ( $n = 3$ ) arteries. The resting and peak CFV were  $0.27 \pm 0.12$  m/s and  $0.65 \pm 0.26$  m/s, respectively. The CFR was  $2.54 \pm 0.75$ . No patient had evidence of acute rejection on biopsy or CAV by coronary angiography.

However, four patients had a CFR  $< 2$ : two of these patients had a low workload and 1 had poor images during SSCE; 1 patient had a normal DSE. All but one patient had a normal stress test; this patient having abnormal wall motion at rest.

**Conclusions:** In this preliminary study, we were able to measure CFV and CFR using DSE and SSCE echocardiography in PHTP. CFR indicated no evidence of CAV in 17/18 tests. The measurement of CFR may have clinical utility as a non-invasive, inexpensive and technically simple tool in assessing CAV following pediatric heart transplantation. However, as there were no cases of rejection nor CAV, further studies are warranted to determine the utility and predictive accuracy of this tool.

#### P-184

##### **Coronary flow in Neonates with Impaired Intrauterine Growth**

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**Introduction:** Subclinical myocardial injury has been reported in newborns with fetal weight below 2 standard deviations for the gestational age. Our aim was to investigate whether impaired intrauterine growth affects cardiac function and coronary flow (CF).

**Methods:** Seventeen newborns with impaired intrauterine growth and fifteen age-matched healthy controls were enrolled in the study. Fetal growth was assessed by fetometry. Doppler velocimetry of the umbilical artery and maternal uterine arteries blood flow was assessed. Cardiac function and left anterior descending artery (LAD) coronary flow were measured by transthoracic Doppler echocardiography at one week of age.

**Results:** The mean growth deviation of the newborns from normal was  $-2.5 \pm 0.2$ . The left ventricular mass and left ventricular shortening fraction was similar in patients and controls. The mean LAD diameter was  $0.99 \pm 0.1$  mm in patients and  $0.8 \pm 0.1$  in controls,  $p = 0.002$ . LAD flow velocity time integral (VTI)/min correlated with left ventricular mass ( $R = 0.46$ ,  $p = 0.0001$ ) and with mitral peak E-wave velocity ( $R = 0.74$ ,  $p < 0.01$ ). Impaired intrauterine growth was associated with increased peak flow velocity in diastole;  $34.5 \pm 4$  cm/s and  $19 \pm 6$  cm/s in controls,  $p = 0.0001$  as well as increased CF;  $37 \pm 7.3$  ml/min and  $8.2 \pm 3.0$  ml/min in controls,  $p = 0.001$ .

**Conclusions:** Coronary flow is significantly increased in neonates with impaired intrauterine growth. The left ventricular mass and systolic and diastolic function remained normal. The clinical significance of the increase of CF is unclear but increased basal flow might lead to a decreased coronary flow reserve.

#### P-185

##### **Echocardiographic evaluation of left ventricular outflow tract hemodynamics in healthy children with anomalous left ventricular band**

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Clinic of Pediatrics, Clinical Center, Nis, Serbia (1); Clinic for cardiovascular diseases – Institute for treatment and rehabilitation, Niska Banja, Serbia (2); Chelsea & Westminster Hospital, London, United Kingdom (3); University Children's Hospital, Belgrade, Serbia (4)

**Basis or Objectives:** The anomalous left ventricular band is considered the most common innocent cardiovascular phenomenon in healthy children. Except for its role in the genesis of

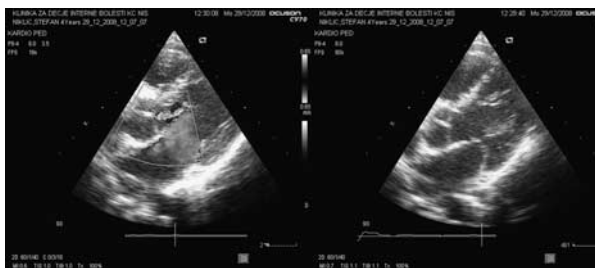


cardiac murmur, ventricle geometry and/or architecture changes, there are still no relevant clinical data confirming its pathologic cardiovascular significance. We hypothesized that high interventricular septal diastolic stretching due to anomalous left ventricular band may cause abnormal subaortic left ventricle outflow tract relaxation and subsequent aortic regurgitation.

**Methods:** To test this, we performed complete transthoracic echocardiography screening on 100 consecutive children with midsystolic vibratory murmur suggestive of anomalous left ventricular band presence. All children were referred to our clinic for the murmur etiology clarification and none of them had history as well clinical finding suggestive for any kind of pediatric disease. Definitely we included in the study 94 children who were found not to have structural cardiac abnormality after the echocardiography examination.

**Results:** According to the position where the anomalous left ventricular band attached the septum, children were conventionally divided into two groups: those with proximal (high third) and those with distal (low or middle third) interventricular septum attachment. Anomalous band was found in 83 children (88.3 percent). Of them, 45 children (54.2 percent) had high (proximal) and 38 children (45.8 percent) had mid or low (distal) anomalous band septal attachment. Aortic regurgitation was found in eight children (17,8%) with proximal anomalous band attachment, of whom 6 children (13,3 percent) had trivial aortic (Ao) regurgitation and 2 children (4,4 percent) mild Ao regurgitation (Fig). Distal anomalous band septal attachment was accompanied by trivial aortic (Ao) regurgitation in only one child (2,6 percent). Ao regurgitation was found more prevalent in children with proximal vs. children with distal anomalous left ventricular band septal attachment ( $p < 0,01$ )

**Conclusions:** Our results show for the first time that anomalous left ventricular band may cause aortic regurgitation in apparently normal hearts. Apart from the need of periodic echocardiographic evaluation, the results of our study might have potential importance in a number of diagnostic dilemmas, especially in cases of aortic valvular regurgitation with absence of aortic root dilatation or other structural aortic valve abnormalities.



#### P-186

**Quantifying tricuspid regurgitation of patients with congenital heart disease routinely by magnetic resonance**  
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**Introduction:** Tricuspid regurgitation is an important clinical entity in congenital heart disease. However, to date tricuspid

regurgitation cannot be quantified reliably. Especially in patients with increased RV pressure tricuspid regurgitation is very difficult to estimate.

Therefore, we sought to define a reliable way to quantify tricuspid regurgitation routinely in congenital heart disease by combining two previously described Cardiovascular Magnetic Resonance (CMR) methods.

**Methods:** 39 consecutive patients (13 female, mean age  $32 \pm 19$  years) with congenital heart disease and assumed tricuspid regurgitation referred to CMR for routine clinical evaluation were included to the study.

Tricuspid regurgitation was quantified by calculating the arithmetic mean of two measurement methods, direct and indirect measurements. Tricuspid regurgitation was measured directly by deducting retrograde from antegrade flow through the tricuspid valve. Retrograde and antegrade flow through the tricuspid valve were independently measured by two CMR phase contrast measurements. These two CMR phase contrast measurements were carefully angled perpendicular to the direction of retrograde or antegrade flow, respectively. Furthermore, each of these two CMR phase contrast measurements had an individual velocity encoding.

Tricuspid regurgitation was measured indirectly using antegrade and retrograde flow through the pulmonary artery and RV stroke volume. Flow through the pulmonary artery was measured by CMR phase contrast flow. RV stroke volume was measured using a standard stack of CMR multi-phase, multi-slice cine sequences. Agreement of indirect and direct measurements was evaluated by Passing-Bablok Regression.

**Results:** There was no significant systematical deviation of both tricuspid regurgitation measures at the zero level (intercept: 0.00). The proportional error per tricuspid regurgitation unit was less than 1% (slope: 1.008, 95% confidence interval: 0.87–1.18). Therefore, a measured tricuspid regurgitation of, for example, 60% can with a probability of 95% in fact be expected between 52% and 71%. Accordingly, a measured tricuspid regurgitation of 10% was with probability of 95% between 9% and 12%.

**Conclusions:** By combining direct and indirect tricuspid regurgitation measurements, tricuspid regurgitation can be quantified in consistent and therefore a clinically relevant way.

High grade tricuspid regurgitation can be excluded even in patients with increased RV pressure.

#### P-187

**Cardiac functions in children with growth hormone deficiency before and during growth hormone replacement therapy**

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**Objectives:** Childhood growth hormone deficiency (GHD) has reduced left ventricular (LV) mass, but impairment of cardiac function has never been documented. The objective was to assess the cardiac effects of GHD and recombinant human growth hormone (rhGH) treatment using conventional echocardiography and tissue Doppler imaging.

**Methods:** In two pharmacological tests, the diagnosis of GHD was based on a peak level of growth hormone less than 10 microgram/liter. Complete two-dimensional, M-mode, pulse wave Doppler echocardiography and pulse wave tissue Doppler imaging were performed in 12 children (six males and six females) with GHD at baseline and  $5.86 \pm 1.61$  month after rhGH therapy. Among nonparametric tests, Wilcoxon test was



used to compare the clinical and cardiac variables before and after rhGH treatment.

**Results:** The rhGH treatment was associated with a significant increase of LV mass index ( $63.8 \pm 27.1$  to  $79.3 \pm 30.3$  g/m<sup>2</sup>;  $p < 0.01$ ) and LV internal dimensions ( $21.4 \pm 2.63$  to  $24.0 \pm 4.13$  mm in systole;  $p = 0.03$  and  $36.5 \pm 3.90$  to  $39.5 \pm 4.94$  mm in diastole;  $p < 0.01$ ). There were statistically difference regarding parameters such as deceleration time of early peak velocity of mitral, isovolumic relaxation time, and myocardial performance index ( $103 \pm 15.4$  to  $139 \pm 21.2$  ms;  $p < 0.01$ ,  $55.5 \pm 9.24$  to  $44.9 \pm 5.44$  ms;  $p < 0.01$ , and  $37.8 \pm 4.46$  to  $69.2 \pm 3.74\%$ ;  $p < 0.01$ , respectively). Before and during rhGH therapy, there were no significant differences regarding fractional shortening of the left ventricle, peak mitral and tricuspid wave velocities with ratios using conventional echocardiography and tissue Doppler imaging.

**Conclusions:** In children, GHD affects heart morphology, by inducing a decrease in cardiac size, but does not modify cardiac function. The rhGH treatment increases cardiac mass, but does not difference parameters of conventional echocardiography and tissue Doppler imaging. It should be mentioned, however, that our series is small and the treatment period is short; other data are thus helpful in confirming the cardiac effects of rhGH therapy in children with GHD.

#### P-188

##### Novel combined 'Cardiac Catheterization-Magnetic Resonance Imaging' (XMR) under dobutamine stress for cardiovascular assessment in children prior to liver transplant

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**Background:** Congenital heart disease is common in children with cholestatic liver disease and has been associated with higher liver transplantation (LT) mortality. Pre-LT cardiovascular assessment to evaluate right heart obstructions and increase in the cardiac index during pharmacological stress can be performed by echocardiography or cardiac catheterization, but at the expense of several limitations and assumptions. We present a novel combined cardiac catheterization-magnetic resonance imaging technique (XMR) using dobutamine-stress for cardiovascular assessment in children considered for LT.

**Methods:** Fourteen children (mean 4.5 years, range 8 months-15 years) with advanced liver disease underwent nineteen XMR investigations. The protocol included (1) anterograde fluoroscopic cardiac catheterization to assess pressures in the right ventricle (RV) and pulmonary arteries (PA) and (2) magnetic-resonance imaging to assess the cardiovascular morphology and measure flows in the great vessels to allow calculation of vascular resistance and cardiac index at rest and during two further stage dobutamine stress (10 µg/kg/min and 20 µg/kg/min).

**Results:** XMR was completed successfully in all nineteen investigations. During maximal dobutamine stress (20 µg/kg/min), there was a significant increase ( $p < 0.05$ ) in the mean heart rate (+54%), RV systolic pressure (+60%), mean PA (+31%), mean femoral artery (FA) (+20%), RV/FA ratio (+25%) and cardiac index (+45%) as shown in Table 1. Patients with baseline elevation of their cardiac index  $> 4.31$  l/min/m<sup>2</sup> (Figure 1) were less likely to further raise the cardiac index above the previously

recommended threshold of  $> 40\%$  (sensitivity 89%, specificity 90%). Of these three were found suitable for LT. The mean X-ray radiation screening time was 8 minutes and radiation dose 2.22 Gy.cm<sup>2</sup> (below 90% of the reported conventional radiation exposure). Seven patients underwent successful LT, two are still listed awaiting LT, three were removed from the waiting list due to their improved liver condition and two died awaiting LT.

**Conclusions:** We propose a new dobutamine-stress XMR protocol for pre-LT cardiovascular assessment, which is feasible, safe and has lower radiation dose than traditional methods. It allows morphology delineation, accurate haemodynamic calculations and evaluation of the cardiac reserve. Previous cardiac index thresholds of a 40% increase during stress may not apply to patients that are already in a hyperdynamic rest state.

Table 1. Haemodynamic changes at rest and at dobutamine stress (10 and 20 µg/kg/min) expressed as mean and (standard deviation).

	Rest	Dobutamine 10 µg/kg/min	Dobutamine 20 µg/kg/min
Heart rate (bpm)	89 (15)	103 (15)	129 (18)
Cardiac index (l/min/m <sup>2</sup> )	4.3 (1.3)	5.5 (1.5)	6.0 (1.4)
RV systolic pressure (mmHg)	40.9 (11.5)	56.3 (21.2)	65.5 (27.9)
Mean PA Pressure (mmHg)	20.3 (4.4)	25.4 (6.2)	26.6 (11.2)
Mean FA Pressure (mmHg)	55.4 (12.7)	66.2 (13.6)	66.6 (15.4)
Ratio	0.52	0.55	0.65
RV/FA	(0.18)	(0.24)	(0.26)

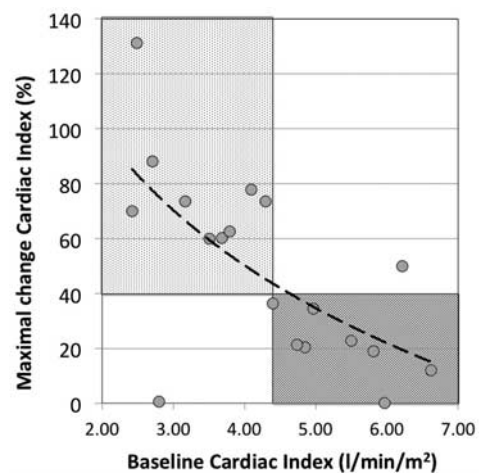


Figure 1. Scatter-plot graph. Maximal change in cardiac index (%) during dobutamine stress compared with the baseline cardiac Index at rest.

#### P-189

##### Detection of early cardiac dysfunction in patients with $\beta$ thalassemia major and thalassemia trait by tissue Doppler echocardiography

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Cardiac complications are the leading cause of death in  $\beta$  Thalassemia Major (TM) patients. The aim of this study was to investigate the impact of iron overload on ventricular functions using the conventional and tissue Doppler imaging (TDI) in patients with TM and compared to the children with thalassemia trait (TT) and healthy controls.

This prospective study included three groups; Group I: 29 patients with TM, Group II: 28 patients with TT, Group III: 29 healthy controls. All the patients and control subjects underwent 2D, M-mode, Doppler and TDI.

No differences were evident between the three groups in age, weight, gender, ejection fraction ( $p > 0.05$ ). Relative wall thickness, left ventricular mass, and left ventricular mass index were found to be higher in the group I ( $p < 0.05$ ). The E/A ratio determined by conventional echocardiography for the right ventricle and left ventricle were significantly lower in TM patients than the other groups ( $E_r/A_r$ :  $1.4 \pm 0.4/1.8 \pm 0.5/1.7 \pm 0.4$ ,  $p < 0.001$ ;  $E_m/A_m$ :  $1.7 \pm 0.3/2 \pm 0.4/1.9 \pm 0.3$ ,  $p < 0.05$ ). Peak late relaxation velocity determined by TDI for the left ventricle ( $A_{dm}$ :  $10 \pm 3.4/6.9 \pm 1.7/7.2 \pm 1.2$ ), interventricular septum ( $A_{di}$ :  $8.4 \pm 2.4/5.9 \pm 1/6.2 \pm 1$ ), and right ventricle ( $A_{dr}$ :  $13.7 \pm 2.9/9.2 \pm 1.8/10.5 \pm 2.3$ ) were significantly higher in TM patients than the TT patients and controls ( $p < 0.001$ ). The early to late relaxation velocity ratio for the left ventricle ( $E_{dm}/A_{dm}$ :  $1.8 \pm 0.9/2.5 \pm 0.4/2.8 \pm 0.6$ ), interventricular septum ( $E_{di}/A_{di}$ :  $1.7 \pm 0.4/2.4 \pm 0.5/2.5 \pm 0.5$ ), and right ventricle ( $E_{dr}/A_{dr}$ :  $1.2 \pm 0.3/1.9 \pm 0.5/1.8 \pm 0.4$ ) were significantly lower in the group I ( $p < 0.001$ ). There was a negative correlation between the ferritin level and E/A ratio for the left ventricle, interventricular septum and right ventricle using the TDI ( $p < 0.05$ ,  $r = -0.378$ ,  $-0.491$ ,  $-0.448$  respectively).

Our results demonstrated that indexes of diastolic function, including TDI, were significantly impaired in TM patients than the TT and healthy children. Conventional echocardiographic techniques have failed to distinguish ventricular functions asymptomatic patients with TM from the patients with TT, and normal controls when global functions were examined. The present study indicates that TDI should be used for screening of TM and TT patient's cardiac functions.

#### P-190

##### Exercise capacity and cardiac reserve in children with corrected pulmonary atresia with intact ventricular septum after univentricular and biventricular repair

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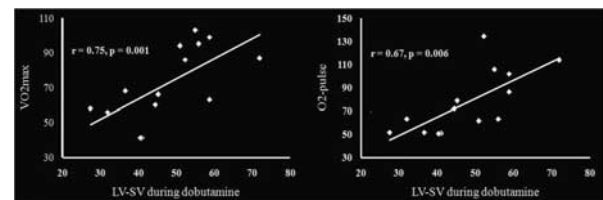
**Introduction:** Management of pulmonary atresia with intact ventricular septum (PAIVS) is challenging and is determined by the degree of the right ventricular hypoplasia after birth. Clinical outcomes of biventricular repair appear favorable to a Fontan operation; however, data on superiority of biventricular repair regarding exercise performance are conflicting. We investigated the response to physical and pharmacological stress in surgically corrected PAIVS patients, with assessment of the presence of myocardial fibrosis.

**Methods:** Sixteen pediatric patients (7 patients after univentricular repair, age  $11.8 \pm 2.6$  years, 7 patients, age and sex matched, after biventricular repair, age  $13.7 \pm 4.5$  years, and 2 patients after one and a half (1.5) ventricular repair underwent a cardiopulmonary

exercise test, dobutamine stress magnetic resonance imaging (MRI) and delayed contrast enhanced MRI. The presence of myocardial fibrosis was assessed and the differences between both groups in response to physical and pharmacological stress were compared between the uni- and biventricular group.

**Results:** The univentricular repair group showed impaired exercise capacity in contrast to normal exercise capacity in the biventricular repair group. At rest, heart rate and left ventricle (LV) functional parameters were not different between both groups. With dobutamine, LV ejection fraction increased in both groups. However, LV stroke volume (SV) increased only in patients after biventricular repair ( $+11.3 \pm 5.0$  ml,  $p < 0.001$ ), but not in patients after univentricular repair ( $-0.04 \pm 3.6$  ml,  $p = .9$ ), HR increase was inadequate in the univentricular repair group. Maximum oxygen consumption ( $VO_2\max$ ) and oxygen-pulse ( $O_2$ -pulse) were strongly correlated with LV-SV during dobutamine (Figure 1) but not to LV-SV at rest. In both groups, no myocardial fibrosis was detected. The results of the 2 patients after 1.5 ventricular repair were comparable to the univentricular repair group.

**Conclusion:** Impaired exercise capacity in children with PAIVS after univentricular repair is related to decreased cardiac reserve due to impaired LV filling during stress, and inadequate chronotropic response. In contrast, pediatric PAIVS patients after biventricular repair show normal exercise capacity and cardiac reserve. These findings support the superiority of biventricular correction of PAIVS over uni- and 1.5 repair during mid-term follow-up.



#### P-191

##### Caution in the Interpretation of Z-Scores in Patients with Pulmonary Atresia with Intact Ventricular Septum

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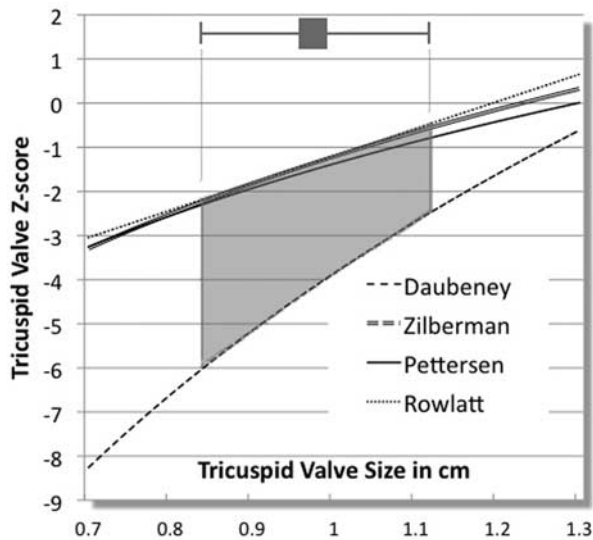
**Introduction:** A common means of expressing how far an observed measurement deviates from the normal is through the use of Z-scores. Z-scores are applied widely in the interpretation of echocardiographic data, and clinical decisions are often based upon them. However, they are extremely sensitive to the properties of the 'normal' population from which they are derived, and also amplify errors in measurement. This study sought to gauge the impact of observer variability on z-scores for a study population with a hypoplastic right heart.

**Methods:** The echocardiograms of 18 consecutive neonates at this institution with pulmonary atresia and intact ventricular septum (PAIVS) were reviewed. Measurements on the tricuspid valve (TV) annulus were made on two separate occasions by three independent observers (two consultants and one registrar).

**Results:** The Interclass Correlation Coefficient (ICC) between the two sets of measurements by each observer was 0.83 (representing good agreement). The ICC between the three observers was 0.77 (representing fair agreement).

The median TV size of the neonates with PAIVS was 0.98 cm. The 95% confidence interval of the mean of the six observations for a valve of this size was 0.84 cm–1.11 cm.

When converted to a z-score for a neonate of median body surface area ( $0.21 \text{ m}^2$ ), the range of z-scores obtained from commonly used data sets ranges from  $-6.0$  to  $-0.6$  (see figure 1).



**Conclusions:** The range of z-scores obtainable from a carefully measured echocardiogram is significant. A z-score of  $> -3.0$  is commonly used as a guide towards biventricular repair in PAIVS, but clinicians should be extremely cautious in their interpretation of z-scores, and should be aware of the wide variation between different published z-score algorithms.

#### P-192

##### Quantitative Evaluation of the Hemodynamics in Fontan Circulation

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**Objectives:** Detecting early ventricular dysfunction and introducing medication therapies with adequate timing are essential to improve long-term outcomes after Fontan procedures. There have been several numerical studies on Fontan circulation discussing the flow energy loss (EL) that reflects the hemodynamic stabilities and which would lead to increase the cardiac workload. However, due to the definition of EL, which requires information of both pressure and velocity distributions, in-vivo clinical measurements of EL have never been reported, and its effects on heart failure were unclear. In the present study, we measured EL in Fontan patients with simultaneous measurements of pressure and velocity, and revealed the influence to the systolic and diastolic functions of the single ventricle.

**Methods:** Catheter exams measuring pressure and velocity simultaneously were performed 1 year after the Fontan procedure in 8 patients. EL was calculated using the averaged pressure and velocity data of 20 cardiac cycles in the superior and inferior vena cava and the bilateral pulmonary arteries. Blood flow amounts were approximated using cross-sectional areas obtained from cineangiograms. The ventricular systolic functions were evaluated with max  $dp/dt$  during the isovolumic systolic phase (Sdpdt), and the ejection fraction (EF) in volumetry; whereas the

diastolic functions were evaluated with max  $-dp/dt$  (Ddpdt), and time constant tau in the isovolumic diastolic phase. The ratio of Ddpdt/Sdpdt was examined to investigate the impaired phase in one cardiac cycle. The echocardiographic Tei index was also examined.

**Results:** EL significantly correlated with Sdpdt ( $r = 0.827$ ), but not with the EF ( $r = 0.472$ ), indicating that high EL reflected afterload increase. EL significantly correlated with tau and tei index ( $r = 0.755$  and  $0.705$ , respectively), but weakly with Ddpdt ( $r = -0.581$ ). EL strongly correlated with Ddpdt/Sdpdt ( $r = 0.886$ ), indicating that EL increased when the diastolic function was impaired, while the systolic function was preserved.

**Conclusions:** EL is a novel and sensitive parameter not only reflecting flow efficiencies in cavopulmonary anastomosis sites but also detecting early heart failure with preserved systolic function with increased afterload and impaired diastolic function of the single ventricle. Quantitative evaluation of the hemodynamics is useful to manage patients with Fontan palliation.

#### P-193

##### Impact of Sport and High Intensity Training on Global and Regional Myocardial Deformation in Elite High School Athletes

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**Background:** Cardiac adaptation to high intensity athletic training is characterized by increases in left ventricular (LV) chamber dimension, wall thickness & mass. The physiologic impact of training on myocardial deformation in highly trained elite high school athletes has not been extensively reported. The purpose of this study was to assess the impact of the type of sport participation & long-term training on parameters of LV systolic & diastolic function including newer modalities of tissue Doppler and myocardial deformation in highly trained high school athletes compared to sedentary controls.

**Methods:** Standard two-dimensional (2D), spectral Doppler, & tissue Doppler were prospectively performed utilizing a GE Vivid 7 to evaluate LV systolic & diastolic function in 78 elite high school athletes (45 males) compared to controls. Longitudinal 2D strain was performed to evaluate 17 regional (apical, mid, & basal segments) & global longitudinal LV myocardial strain. The impact of the type of sport & training (figure skating, ice hockey, soccer) on LV systolic & diastolic function were assessed.

**Results:** Data are summarized in the table. Heart rate, LV dimensions, and LV mass were significantly different in athletes compared to healthy controls ( $p < 0.001$ ). Traditional measures of LV systolic and diastolic function as well as tissue Doppler and global longitudinal 2D strain were not different between groups. However, regional differences in strain were identified, with apical deformation increased relative to basal function in athletes compared to controls ( $p < 0.001$ ). Similar regional changes were seen in all athletes, the most prominent differences in skaters and hockey players.

**Conclusions:** While global longitudinal myocardial velocity and deformation were not significantly different in athletes compared to healthy controls, regionally increased apical deformation compared to basal function was consistently demonstrated in all athletic groups. Ongoing assessment of radial and circumferential deformation as well as LV torsion may add novel insights into whether this represents a compensatory adaptation or augmented contractile reserve with training in athletes. This information

may also assist in distinguishing athletic from myopathic increases in LV mass.

	Athletes	Controls	
N	78	25	
Age	15.9 +/- 1.3	15.6 +/- 1.9	p = NS
BSA	1.8 +/- 0.2	1.7 +/- 0.2	p = NS
HR	62 +/- 11	71 +/- 13	p < 0.001
LV Mass	147 +/- 34	131 +/- 38	p < 0.001
LV EDD	5.1 +/- 0.4	4.8 +/- 0.5	p < 0.001
LV ESD	3.2 +/- 0.4	3.0 +/- 0.3	p < 0.01
LV EF %	61 +/- 6	60 +/- 5	p = NS
TDI Septal E	0.15 +/- 0.02	0.14 +/- 0.02	p = NS
TDI Septal A	0.06 +/- 0.01	0.06 +/- 0.01	p = NS
TDI Septal S	0.09 +/- 0.01	0.09 +/- 0.01	p = NS
TDI Mitral E	0.19 +/- 0.03	0.20 +/- 0.03	p = NS
TDI Mitral S	0.11 +/- 0.02	0.11 +/- 0.03	p = NS
Global 2D Strain	-20.3 +/- 1.8	-20.7 +/- 1.9	p = NS
Apical 2D Strain	-21.9 +/- 3.4	-20.0 +/- 2.8	p < 0.001
Mid 2D Strain	-20.1 +/- 1.7	-20.4 +/- 2.0	p = NS
Basal 2D Strain	-19.0 +/- 1.8	-21.9 +/- 2.7	p < 0.001

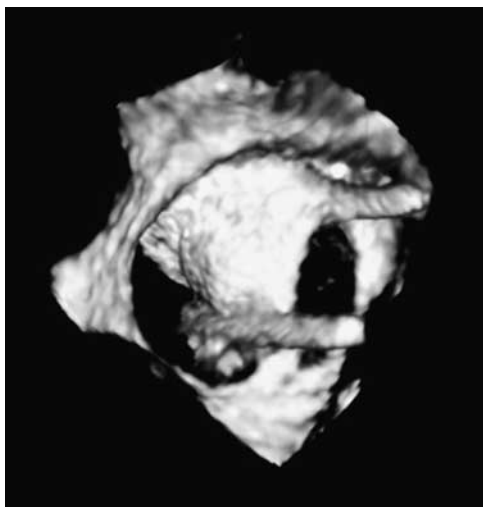
### P-194

#### Value of Real – Time Three – Dimensional Transesophageal Echocardiography in Assessment and Percutaneous Closure of Multiple Atrial Septal Defects

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**Introduction:** Evaluation and percutaneous closure of multiple or multifenestrated atrial septal defects (ASDs) can often be challenging under conventional two – dimensional transesophageal echocardiographic (2DTEE) guidance. Real – Time Three Dimensional Transesophageal Echocardiography (RT3DTEE) is a recently introduced technique, capable of real-time three-dimensional imaging with no need for multiple – beat acquisition with ECG gating and off-line processing. It is particularly useful for guidance of percutaneous procedures, when detailed information regarding number, size, shape and spatial relationships of multiple ASDs is required and when locating guidewires and catheters and assessing multiple device position are essential.



**Methods:** We performed RT3DTEE in 8 patients (5 females, age range 12–63 years) undergoing cardiac catheterization for percutaneous closure of ASDs. Of them, 6 had a previous transthoracic echocardiographic diagnosis of multiple or multifenestrated ASDs.

**Results:** RT3DTEE provided detailed en-face views of the atrial septum on both sides and thorough information regarding size, shape and spatial arrangement of the defects. It showed a multifenestrated/cribrous-type defect in 2 patients, and multiple sizeable ASDs in the remaining 6. The atrial septum appeared aneurysmal in 5 patients. Three patients, due to deficient margins and excessive size of the defects, were deemed unsuitable for percutaneous treatment, and referred for surgical closure. Surgical exploration confirmed RT3DTEE findings in both. Five patients underwent successful transcatheter closure of the ASDs, in 3 cases with a single large device, and with two devices in 2 cases. RT3DTEE enabled accurate location of guidewires, sizing balloons and catheters when crossing the defects, ensuring correct choice and placement of the devices. In the cases where two devices were used, RT3DTEE provided information on their arrangement and relationship with surrounding structures. **Conclusions:** Our preliminary data show that RT3DTEE is a feasible and accurate technique that provides reliable information and guidance during transcatheter closure of multiple ASDs, overcoming many of the limitations of 2DTEE in managing this complex type of defects.

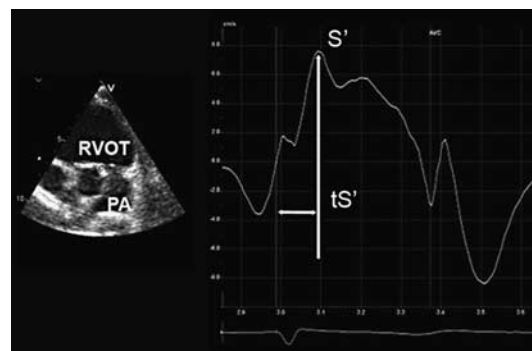
### P-195

#### Tissue Doppler imaging in healthy children: normal systolic velocities, timings, and time differences in left ventricle and right ventricle

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**Introduction:** Tissue Doppler imaging (TDI) has recently emerged as an important diagnostic tool to assess myocardial velocities and timings of peak systolic velocities at the right (RV) and left ventricle (LV) and within the ventricles. However, in children, normal age-related values of peak systolic velocities, time to peak systolic velocities and intra-ventricular time differences within the LV and RV remain largely unknown. Normal age-related values of peak systolic velocities in children could serve as a reference for future studies on global and regional myocardial performance in pediatric patients. Reference values on intra-ventricular time differences could help to refine therapeutic strategies in pediatric patients, including cardiac resynchronisation therapy (CRT).



The objectives of this study were to assess peak systolic velocities, timings of peak systolic velocities and intra-ventricular time differences in the LV and RV of healthy children with TDI and to provide age-related normal values.



**Methods:** Consecutive healthy children (0–18 years) underwent transthoracic echocardiography. TDI data was acquired at the conventional apical 4-chamber view and at a dedicated apical RV outflow tract (RVOT) view. Peak systolic velocity (S') and time to peak systolic velocity (tS') were assessed at the basal segments of the LV lateral wall (LVlat), inter-ventricular septum (IVS), RV free wall (RVFW) and at the RVOT. Time differences in peak systolic velocities within the RV and LV were calculated. Regression analysis was performed to assess the age-dependency of the observed TDI data.

**Results:** A total of 123 children were included. The observed TDI values are summarized in Table 1. Within the RV of healthy children, a significant time delay was observed. At all regions, S' significantly increased with age (LVlat:r = 0.75, p < 0.01, IVS:r = 0.54 p < 0.01, RVFW: r = 0.42, p < 0.01, RVOT:r = 0.38, p < 0.01). Furthermore, tS' related significantly with age at all regions (LVlat:r = 0.57, p < 0.01, IVS:r = 0.41, p < 0.01, RVFW:r = 0.58, p < 0.01, RVOT:r = 0.70, p < 0.01). The observed intra-ventricular time differences did not correlate with age. **Conclusions:** In healthy children, there is a significant intra-ventricular time delay within the RV. TDI-derived peak systolic velocities and timings are significantly related to age in healthy children. However, intra-ventricular time differences are not related to age in healthy children.

Table 1. TDI-derived values of peak systolic velocities, timings and time differences.

Peak systolic velocity (cm/s)	
<i>Left ventricle</i>	
LVlat	6.3 (5.1–7.9)
IVS	6.0 (5.4–6.7)
<i>Right ventricle</i>	
RVFW	10.2 (8.9–11.3)
RVOT	7.2 (6.0–8.2)
Time to peak systolic velocity (ms)	
<i>Left ventricle</i>	
LVlat	101 (91–112)
IVS	114 (100–128)
<i>Right ventricle</i>	
RVFW	177 (157–194)
RVOT	100 (88–113)
Time differences (ms)	
<i>Left ventricle</i>	
LVlat – IVS	10 (0–20)
<i>Right ventricle</i>	
IVS – RVFW	62 (45–75)
RVOT – RVFW	74 (59–93)

Values are presented as median and inter-quartile range.

IVS: inter-ventricular septum, LVlat: left ventricular lateral wall

RVFW: right ventricular free wall, RVOT: right ventricular outflow tract

## P-196

### Branch Pulmonary Arterial Phase-Contrast MRI to detect pulmonary venous baffle obstruction after atrial switch procedure

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**Introduction:** It has been shown that pulmonary vein stenosis results in reduced ipsilateral arterial forward flow with diastolic

reversal and contralateral diastolic forward flow (Roman, 2005). However, pulmonary venous baffle stenosis occurs uniquely as a complication of a previous atrial switch procedure for transposition. The aim of this study was to investigate these arterial flow patterns in this setting.

**Methods:** We retrospectively reviewed 2006–2009 data. Patients with previous atrial switch procedure & without residual shunts who had undergone flow evaluation in the main (MPA), right (RPA) and left (LPA) pulmonary arteries using Phase-Contrast MRI (PC-MR) were included. PC-MR data was correlated with the severity of pulmonary vein stenosis as defined on contrast-enhanced MR angiography (diameter reduction: mild: 25–50%; moderate: 50–75% and severe: >75%).

**Results:** 9 patients met the inclusion criteria. One patient had a severe left pulmonary venous baffle stenosis and two patients had a mild left stenosis. No patient had right pulmonary venous obstruction. One patient was not evaluated for continuous or reversed arterial flow due to pulmonary regurgitation. Only one of the sixteen branch pulmonary arteries showed any significant continuous diastolic forward flow and this was in the RPA of the patient with a severe left pulmonary venous baffle stenosis. Similarly, the LPA in this patient was the only pulmonary artery to show significant flow reversal in diastole (Figure 1). Three patients showed >65% of flow distributed to the RPA, all of whom had left pulmonary venous baffle stenosis. There was a significant correlation of the degree of pulmonary baffle stenosis with RPA/LPA flow ratio (Pearson's Coefficient = 0.74 p = 0.02) but not with RPA/LPA cross-sectional area ratio (-0.26, p = 0.5).

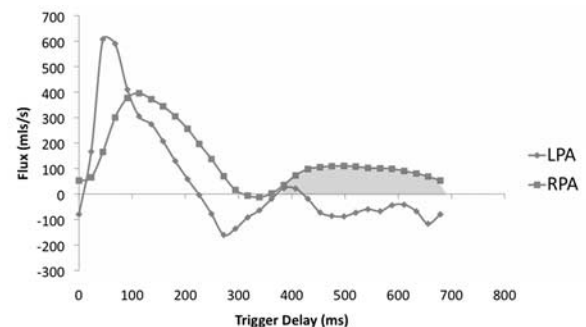


Figure 1: Flow Patterns in a patient with severe left pulmonary venous baffle stenosis. Approximately 40% reversal of flow is seen in the LPA in diastole (negative flux): This correlates with the systolic forward flow in the RPA (shaded). The overall distribution of flow RPA:LPA is 78%:22%.

**Conclusions:** Redistribution of pulmonary arterial blood flow may make conventional echocardiographic measures of pulmonary venous stenosis difficult to interpret. Arterial flow redistribution appears to be a sensitive marker for pulmonary venous baffle stenosis. In the absence of pulmonary regurgitation, reversed diastolic flow in the ipsilateral and continuous in the contralateral pulmonary artery appears specific for severe obstruction. Branch pulmonary artery PC-MR should be performed routinely for patients who have had an atrial switch for transposition.

## P-197

### Is exercise capacity of adults with repaired tetralogy of Fallot affected by type of repair?

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**Purpose:** Adult patients with repaired tetralogy of Fallot (rToF) have impaired exercise tolerance. The primary objective of this

study was to determine whether different types of procedures affect the exercise capacity of these patients.

**Methods:** A retrospective analysis of the cardiopulmonary exercise testing results of a cohort of 33 patients (30% females) operated for tetralogy of Fallot was performed. Patients were divided into two groups, according to the type of reparative procedure: group A (n = 18) operated with right ventricle to pulmonary artery conduit and group B (n = 15) submitted to trans-atrial, trans-pulmonary repair. All patients underwent cardiopulmonary exercise testing. We measured peak oxygen consumption (peak VO<sub>2</sub>), the slope of ventilation per unit of carbon dioxide production (VE/VCO<sub>2</sub>) and calculation of peak VO<sub>2</sub> to predicted VO<sub>2</sub> according to age and sex (VO<sub>2</sub>pred%) and oxygen pulse (O<sub>2</sub> pulse). Right ventricular end diastolic diameter (RVEDD) and right ventricular systolic pressure (RVSP) were measured with transthoracic echocardiography. NYHA functional class was also determined for all patients.

**Results:** Our results are depicted in the following table. 26 out of the 33 patients were classified as NYHA functional class I. (78.8%). None of the parameters measured (Table) differed statistically significantly.

**Conclusion:** Type of surgical repair does not seem to affect the exercise ability of patients with rToF. Patients of both groups had impaired exercise capacity, while most of them were classified as NYHA functional class I. In order to estimate the true exercise capacity and prevent fatal events, it is very important to submit these patients to close follow up and evaluate them with cardiopulmonary exercise testing.

Table: Cardiopulmonary testing results. Data are presented as mean ± SD. CPX: cardiopulmonary testing

Data	Total No of Pts n = 33	Group A n = 18	Group B n = 15	p value
Age at CPX (years)	17.6 ± 8	17.7 ± 7.5	17.6 ± 8.9	0.74
Age at repair (years)	3 ± 3.1	3 ± 2.3	4.5 ± 3.9	0.16
VO <sub>2</sub> peak (ml/Kg/min)	29.4 ± 9.7	29.3 ± 8.9	28.8 ± 9.2	0.59
VO <sub>2</sub> peak (%predicted)	69.1 ± 29.8	71.2 ± 30.8	69.1 ± 8.8	0.61
VE/VCO <sub>2</sub>	33 ± 8.7	33.5 ± 6.5	33 ± 10.9	0.68
O <sub>2</sub> pulse	8.2 ± 2.6	10 ± 3	6.9 ± 2	0.40
EDDRV (mm)	32 ± 7.9	30 ± 8	37 ± 7	0.39
RVSP (mmHg)	41 ± 16.3	45 ± 18.3	37 ± 15.1	0.24

#### P-198

##### Is New York Heart Association classification an appropriate tool for the functional evaluation of patients after total repair of Tetralogy of Fallot?

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**Purpose:** The classification of functional status of patients with repaired tetralogy of Fallot (rToF) often relies on New York Heart Association classification, NYHA. However, often, these patients adapt to long-standing limited exercise and underreport symptoms. Cardiopulmonary exercise (CPX) parameters have been shown to be independent predictors of cardiac disease severity. The purpose of this study was to evaluate the relationship between NYHA classification and exercise capacity and to assess the validity of NYHA as a method of functional evaluation of patients with rToF.

**Methods:** Thirty-three consecutive patients with rToF were evaluated during follow-up from December 2000 to December 2009. These patients were divided into three groups according to their NYHA class of functionality: Group A (NYHA I, n = 17), group B (NYHA II, n = 12) and group C (NYHA III, n = 4).

Subsequently CPX was performed with measurement of peak VO<sub>2</sub> and the slope of VE/VCO<sub>2</sub> and calculation of peak VO<sub>2</sub> to predicted VO<sub>2</sub> according to age and sex (VO<sub>2</sub>pred%) and oxygen pulse (O<sub>2</sub> pulse). Data from all groups were cross-matched (three pairs) and retrospectively analyzed to determine if a correlation exists between NYHA class and exercise capacity.

**Results:** Demographic parameters and CPX results are depicted in table.

Parameters	Total no of pts n = 33	Group A NYHA 1 n = 17	Group B NYHA 2 n = 12	Group C NYHA 3 n = 4
Age at CPX (years)	19.9 ± 8	19.4 ± 8.3	18.9 ± 7.3	24.8 ± 9.7
VO <sub>2</sub> peak (ml/Kg/min)	28.9 ± 9	31.4 ± 9.1	24.1 ± 7.2	24.5 ± 6.9
VO <sub>2</sub> peak (%predicted)	74.1 ± 29.8	85.7 ± 29.4	59.2 ± 23.8	56.6 ± 26.5
VE/VCO <sub>2</sub>	33.9 ± 8.7	31.1 ± 6.5	37.3 ± 11.1	35.5 ± 5.7
O <sub>2</sub> pulse	10.8 ± 4.7	12.1 ± 5.1	14.5 ± 3	6.6 ± 3.6

Group C patients had significantly reduced VO<sub>2</sub> pred% compared with group A patients (p = .02) and O<sub>2</sub> pulse compared with group B patients (p = .03). All other parameters determined were not statistically different among the three groups.

**Conclusion:** Although NYHA classification maintains its value as a method for functional evaluation in patients with rToF and moderate cardiac disease (NYHA III), it has not the discriminatory power to functionally stratify patients with rToF and mild cardiac disease (NYHA I – II). This last group of patients should be closely evaluated with CPX in order to assess the true exercise capacity and prevent fatal events.

#### P-199

##### Right Ventricular Mechanical Dyssynchrony is Associated with Tricuspid Regurgitation in Children with Hypoplastic Left Heart Syndrome

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**Background:** The presence of tricuspid regurgitation (TR) is an important risk factor for adverse outcome in hypoplastic left heart syndrome (HLHS). We have previously shown that right ventricular (RV) mechanical dyssynchrony is common in HLHS, but it is unknown whether it is associated with TR in HLHS. The aim of this study was to investigate RV mechanical dyssynchrony in relation to the presence of TR in children with HLHS.

**Methods:** Children (0–18 years) with HLHS at all stages of surgical palliation were identified from the echo database (2004–2010). 2D echocardiograms were reviewed by 2 observers. RV dyssynchrony was defined by RV intraventricular delay, measured as difference in time to peak systolic tissue velocity between basal RV lateral wall and septum in the apical 4 chamber view equivalent. Patients were classified into 2 groups depending on presence or absence of TR, and results compared using Mann-Whitney test or ANOVA.

**Results:** Forty-eight patients were identified, median (IQR) age 0.59 (0.12–2.94) years, 15% had no previous surgery, 27% were at stage 1, 32% at stage 2 and 26% at stage 3 of palliation. 35 (73%) had TR; 23 (48%) had mild, 2 (4%) moderate and 10 (21%) severe TR. There was no significant difference in RV function between those with and without TR by either subjective assessment, RV fractional area change (37.5 ± 10.8 vs 38.1 ± 6.5%, p = 0.818) or peak RV longitudinal systolic tissue velocity (1.18 ± 0.51 vs 1.28 ± 0.74 cm/s, p = 0.8). RV intraventricular delay was significantly higher in children with TR vs those without (mean 117 ± 159 vs 36 ± 39 msec, p = 0.03). RV intraventricular delay did not change significantly with stage of palliation; 93 ± 136 msec in patients without surgery, 47 ± 35 msec

following stage 1,  $108 \pm 106$  msec following stage 2,  $151 \pm 242$  msec following stage 3 palliation ( $p = 0.3$ ).

**Conclusion:** RV mechanical dyssynchrony is worse in children with HLHS who have TR. This has important implications for further investigation into risk factors, prognosis and possibility of treatment with resynchronization therapy.

#### P-200

##### **Optimizing ECG-triggering during cardiovascular magnetic resonance and its impact on blood flow quantification in patients with congenital heart disease**

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**Background:** Optimal ECG-triggering is of paramount importance for correct blood flow quantification during cardiovascular magnetic resonance (CMR). However, optimal ECG-triggering and therefore blood flow quantification is impaired in many patients with congenital heart disease (CHD) due to complex QRS patterns. Therefore, a new ECG-trigger algorithm was developed to address triggering problems due to complex QRS-patterns.

The aim of this study was to test this new ECG-trigger algorithm in routine patients with CHD and its impact on blood flow quantification.

**Methods:** 35 consecutive routine patients with CHD undergoing CMR were included in the study. (40% Fallot's Tetralogy, 20% aortic arch, 14% TGA, 26% others; age  $26 \pm 11$  yrs)

In all patients blood flow in the ascending aorta was quantified using the standard ECG-trigger algorithm and the new ECG-trigger algorithm in random order. Blood flow quantified using the standard or new ECG-trigger algorithm was compared by Bland-Altman analysis.

Four blinded investigators evaluated the vector clouds and trigger points of both ECG-trigger methods. Evaluation criteria were false positive and false negative triggered QRS-complexes (specificity and sensitivity), and accuracy of detection. Accuracy of detection was defined as time scatter of the trigger around the correct trigger point. **Results:** Blood flow quantification using the standard or new ECG-trigger algorithm differed more than 5% in 31% of the cases.

Specificity, sensitivity, and accuracy ( $p = 0,028$ ) of detection significantly increased using the new ECG-trigger algorithm compared to the standard ECG-trigger algorithm.

**Conclusion:** Our results show that optimizing ECG-triggering during CMR has a large impact on blood flow quantification in approximately 1/3 of routine patients with CHD. We furthermore suggest that incorrect ECG-triggering is a major source of error in blood flow quantification of many patients with CHD undergoing CMR.

#### P-201

##### **Serial follow-up of biventricular function, contractile reserve, exercise capacity, and NT-proBNP measurements in repaired tetralogy of Fallot**

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**Introduction:** Predicting the course of right ventricular (RV) remodelling after repair of tetralogy of Fallot (TOF) is difficult and serial follow-up studies are limited. Our aim was 1) to study the course of biventricular size, function, and contractile reserve in patients with repaired TOF, in relation to exercise capacity and NT-proBNP levels and 2) to establish guidelines for cardiovascular magnetic resonance (CMR) imaging intervals.

**Methods:** We performed serial follow-up in 36 TOF patients ( $15.1 \pm 4.9$  years at baseline; interval 5.1 (4.1-7.4) years) using CMR imaging at rest and during dobutamine stress, exercise testing, and NT-proBNP measurements. Subgroup analysis was performed, based on RV end-diastolic volume (RVEDV) at follow-up: subgroup I:  $<150$  ml/m<sup>2</sup>, subgroup II:  $\geq 150$  ml/m<sup>2</sup>. Nine patients underwent pulmonary valve replacement (PVR) during follow-up and were analyzed as third subgroup.

**Results:** In subgroup II, RV volumes and pulmonary regurgitation (PR) increased significantly during follow-up; this did not occur in subgroup I. Peak oxygen uptake (VO<sub>2</sub> max.) tended to decrease in subgroup II; other exercise parameters did not. Biventricular function, contractile reserve, and NT-proBNP levels remained stable in both subgroups and were not different between all 3 subgroups at follow-up. RV ejection fraction and left ventricular contractile reserve improved after PVR. A lower RV contractile reserve at baseline correlated with a larger RVEDV at follow-up and with an interstudy decrease in VO<sub>2</sub> max.

**Conclusions:** In patients with an RVEDV  $< 150$  ml/m<sup>2</sup>, no changes in ventricular function and clinical state occur during 5 year follow-up. We believe a conservative approach towards CMR imaging intervals and PVR is justified in these patients. Patients with an RVEDV  $\geq 150$  ml/m<sup>2</sup> should be monitored closely, since RV volumes and PR increase over time. VO<sub>2</sub> max. tended to decrease in these patients, but biventricular function, contractile reserve, and NT-proBNP remained stable, and did not differ from results in the other subgroups. In the absence of clinical deterioration, a 2-3 year interval between CMR imaging studies seems justified in patients with an RVEDV  $\geq 150$  ml/m<sup>2</sup>. CMR stress imaging might be of additional value in the follow-up after TOF repair.

#### P-202

##### **Right ventricular function is impaired one week after corrective surgery of an atrial septal defect**

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**Introduction:** Before surgery the heart of an atrial septal defect (ASD) patient is characterized by an increased volume load of the right ventricle. After closure of an ASD long-term results show a small impairment in right ventricular function. The time course of ventricular adaptation shortly after surgery is as yet unknown. Consequently the possible role of an earlier impairment of ventricular function in the development of long term dysfunction is indistinct. Novel echocardiographic Tissue Doppler Imaging (TDI) enables sensitive quantitative assessment of ventricular function. Using TDI we studied ventricular function before and shortly after surgical ASD closure.

**Methods:** The study population consisted of 26 children with an ASD. (0-17 years). Complete echocardiographic studies were

performed before surgery and at discharge ( $6 \pm 1$  days after surgery). Special attention was paid to systolic and diastolic left and right ventricular function, including TDI. Age matched controls ( $N = 41$ ) were used for comparison.

**Results:** Right ventricular (RV) systolic function was impaired at discharge, as assessed by both Tricuspid Annular Plane Systolic Excursion (patients versus controls  $10 \pm 2$  mm versus  $19 \pm 4$  mm;  $P < 0.001$ ) and systolic TDI measurements of the RV free wall (patients versus controls  $5.5 \pm 1.9$  cm/s versus  $13.1 \pm 2.7$  cm/s;  $P < 0.001$ ). Right ventricular diastolic function was impaired at discharge as well, as assessed in patients versus controls by  $E'$  ( $6.5 \pm 2.5$  cm/s versus  $17.7 \pm 3.7$  cm/s),  $A'$  ( $4.6 \pm 1.9$  cm/s versus  $10.5 \pm 1.9$  cm/s) and  $E/E'$  ( $12.3 \pm 9.2$  versus  $4.3 \pm 1.5$ ; all  $P < 0.001$ ) of the RV basal free wall.

In contrast, left ventricular (LV) systolic and diastolic function was normal in patients compared to controls at discharge, as assessed by LV basal free wall  $S'$  ( $6.6 \pm 1.8$  cm/s versus  $6.5 \pm 1.6$  cm/s),  $A'$  ( $4.7 \pm 2.1$  cm/s versus  $5.7 \pm 1.7$  cm/s) and  $E/E'$  ( $9.4 \pm 3.6$  versus  $8.1 \pm 2.7$ ; all NS).

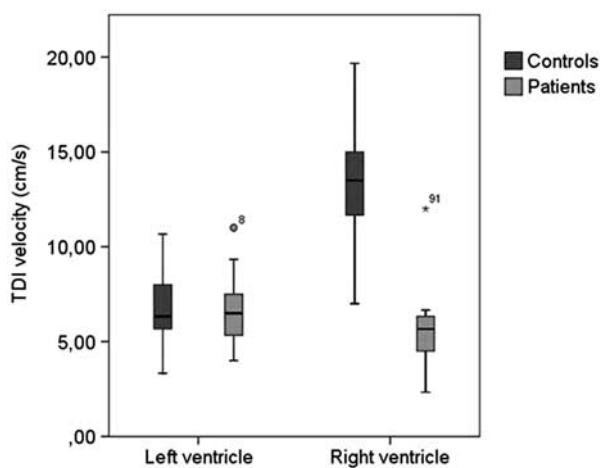


Figure 1. Boxplot presenting the systolic TDI velocity of the LV and RV in patients at discharge versus controls.

**Conclusions:** In contrast to left ventricular function, which was normal at discharge after surgical ASD closure, right ventricular systolic and diastolic function were impaired at discharge. These findings may be due to the preoperative RV volume load, the effects of surgery or the use of cardiopulmonary bypass. The relation between these short-term changes and long-term outcome has further to be elucidated.

#### P-203

##### Shortly after corrective surgery of a ventricular septal defect right ventricular function is impaired and the level of impairment is correlated with cardiopulmonary bypass time

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**Introduction:** Little is known about the changes in ventricular function shortly after surgery of a ventricular septal defect (VSD). Additionally

the influence of load changes, cardiopulmonary bypass and surgery on ventricular adaptation is unclear. Tissue Doppler Imaging (TDI) allows for sensitive quantification of ventricular function. We studied the ventricular function shortly after correction of a VSD.

**Methods:** Complete echocardiographic studies of the left and right ventricle were performed in 42 children with a VSD. (0–17 years) and 20 age-matched controls. Systolic ventricular function was assessed using conventional echocardiography, including ejection fraction (EF) and Tricuspid Annular Plane Systolic Excursion (TAPSE), and systolic TDI ( $S'$ ). Diastolic function was assessed using TDI ( $E'$ ,  $A'$  and  $E/E'$ ). Studies were performed preoperatively, one day after surgical correction and at discharge.

**Results:** In the left ventricle (LV), EF and LV free wall  $S'$  were comparable to controls preoperatively and one day postoperatively. Both parameters were still comparable to controls at discharge ( $51 \pm 9\%$  versus  $53 \pm 6\%$  and  $5.6 \pm 1.7$  cm/s versus  $6.3 \pm 1.6$  cm/s respectively; NS). LV basal free wall  $E'$  and  $A'$  were normal compared to controls preoperatively. One day postoperatively both parameters were slightly impaired but restored to control values at discharge.

In the right ventricle (RV), TAPSE was slightly lower than controls preoperatively and decreased postoperatively. At discharge TAPSE remained impaired (patients versus controls  $8 \pm 2$  mm versus  $17 \pm 4$  mm;  $p < 0.001$ ). All RV basal free wall TDI velocities were comparable to controls preoperatively and decreased one day postoperatively. At discharge RV function was still impaired compared to controls;  $S'$  ( $5.0 \pm 1.6$  cm/s versus  $12.1 \pm 2.5$  cm/s),  $E'$  ( $7.4 \pm 2.8$  cm/s versus  $18.3 \pm 4.9$  cm/s),  $A'$  ( $4.6 \pm 1.9$  cm/s versus  $10.5 \pm 2.6$  cm/s) and  $E/E'$  ( $13.6 \pm 7.8$  versus  $4.7 \pm 1.9$ ; all  $P < 0.001$ ). Additionally, longer aortic cross-clamp time correlated with larger reductions of the RV  $E'$  and  $A'$  one day postoperatively.

**Conclusions:** In contrast to left ventricular function, which differs little from normal values during short term follow up after VSD correction, right ventricular systolic and diastolic function deteriorated postoperatively and remained impaired at discharge. The correlation between aortic cross-clamp time and TDI velocities could imply cardiopulmonary bypass adds to the deterioration of RV diastolic function. Future research is necessary to further clarify especially right ventricular adaptation after surgery.

#### P-204

##### Ventricular function is impaired shortly after corrective surgery of a congenital heart defect

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**Introduction:** Although survival rates are high for corrective surgery in patients with a congenital heart defect (CHD), long term studies demonstrate a varying degree of cardiac dysfunction. Thus far the time course and influencing factors of ventricular adaptation after surgery of a CHD are mostly unknown. Tissue Doppler Imaging (TDI) enables sensitive assessment of ventricular function. In this study we evaluated the adaptation of ventricular function shortly after surgical CHD correction.

**Methods:** We included 141 CHD patients (0–17 years) undergoing biventricular surgical correction. Left and right ventricular function was assessed using echocardiography. Systolic function was assessed using conventional echocardiographic measurements (ejection



fraction (EF) and Tricuspid Annular Plane Systolic Excursion (TAPSE)) and systolic TDI (S'). Diastolic function was assessed using diastolic TDI parameters (E', A' and E/E'). Studies were performed preoperatively, one day postoperatively and at discharge (9 ± 5 days after surgery) in patients and in 41 age-matched controls.

TDI Velocities (cm/s)	Controls	Patients at discharge	P-value
<b>LV</b>			
S'	6.5 ± 1.6	5.7 ± 2.0	0.018
E'	14.5 ± 3.8	9.8 ± 3.9	0.001
A'	5.7 ± 1.7	5.0 ± 2.2	0.078
E/E'	8.1 ± 2.7	12.2 ± 6.4	0.001
<b>RV</b>			
S'	13.1 ± 2.7	5.2 ± 1.7	0.001
E'	17.7 ± 3.7	7.3 ± 2.5	0.001
A'	10.5 ± 1.9	4.2 ± 1.9	0.001
E/E'	6.4 ± 1.7	12.5 ± 6.8	0.001

**Results:** In the left ventricle (LV), EF was slightly enhanced preoperatively but decreased after surgery. EF was normal in patients compared to controls at discharge (52 ± 9% versus 51 ± 5%; NS). LV free wall S', E' and E/E', were comparable to controls preoperatively and deteriorated one day after surgery. These parameters remained impaired at discharge (Table 1).

In the right ventricle (RV), TAPSE was slightly impaired preoperatively and deteriorated after surgery. At discharge TAPSE remained impaired (patients versus controls 9 ± 3 mm versus 19 ± 4 mm;  $p < 0.001$ ). RV basal free wall TDI S', E', A' and E/E' were comparable to controls preoperatively and deteriorated one day after surgery. At discharge these parameters remained impaired (table 1). Additionally, a longer aortic cross clamp time was associated with a larger reduction in the S' and E' velocity of the LV and RV at discharge ( $p < 0.05$ ).

**Conclusions:** TAPSE and TDI measurements suggest systolic and diastolic right and left ventricular function are impaired directly after corrective surgery of a CHD and remain impaired up to discharge. Both techniques were more sensitive than other conventional parameters. A negative association was shown between several TDI velocities and aortic cross clamp time. Persistence of these changes in long term follow up has to be evaluated in future research.

#### P-205

##### **Decreased global myocardial function in infants with hypoplastic left heart syndrome (HLHS) is associated with missed intrauterine diagnosis, longer operative times and lower postoperative systemic blood pressure values**

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**Introduction:** Continuous improvements in surgical and perioperative care have led to increasing survival of infants with HLHS. However, the morbidity rates and hospitalisation times remain high especially when associated with poor cardiac function. We evaluated in uni-/multivariate analysis the clinical and operative factors (eg. type of shunt) association with detailed global functional analysis in HLHS infants before and after the Norwood procedure.

**Methods:** We retrospectively reviewed population based cohort of 55 HLHS infants who underwent the Norwood procedure between January 2003 and March 2010 in our institute. Global right ventricular fractional area change, ejection fraction, myocardial velocity and strain rate were analysed from apical 4-chamber view using Velocity-Vector-Imaging technique (Syngo USWP 3.0, Siemens) both before and after the procedure. Postoperative functional analysis was recorded during the pre-Glenn catheterisation. Intra- and interobserver correlations as well as the correlations between cardiac functional methods were good ( $R > 0.7$ ,  $p < 0.05$ , respectively).

**Results:** Intrauterine diagnosis improved all myocardial functional parameters preoperatively in multivariate analysis ( $p < 0.05$ ). Postoperatively, lower systolic and diastolic blood pressures had negative impact on global myocardial function in multivariate analysis ( $p < 0.05$ ). Interestingly, although the systolic and diastolic blood pressures were significantly lower (82.0 ± 12.8 mmHg vs 93.5 ± 11.9 mmHg,  $p = 0.01$ ; 43.1 ± 10.6 mmHg vs 53.8 ± 8.9 mmHg,  $p = 0.003$ ) in the group of patients with BT-shunt ( $n = 22$ ), the type of the shunt did not have independent impact on global cardiac function. This was related to the fact that the aortic cross clamping and perfusion times were in turn longer in the RV-PA shunt ( $n = 33$ ) group (62.4 ± 22.9 min vs 44.3 ± 25.6 min,  $p = 0.01$ ; 174.9 ± 37.5 min vs 148.2 ± 47.3 min,  $p = 0.03$ , respectively) and associated to the decreased function in univariate analysis ( $p < 0.05$ ).

**Conclusions:** Intrauterine diagnosis is associated with improved global cardiac function in newborn HLHS patients. Lower systemic blood pressure, risk increased especially in the group of BT-shunt patients, is associated with decreased global cardiac function after the Norwood procedure. Similarly, longer operative times, related more commonly to complex procedure of RV-PA shunt, have negative influence on postoperative myocardial function. The long-term impact of these cardiac functional risk factors needs to be defined.

#### P-206

##### **Obesity is responsible for increased blood pressure, left ventricular thickness and tension leading to early impairment of diastolic function**

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**Introduction:** Obesity has been shown to lead to hypertension and increased left ventricular (LV) mass. The aim of this study was to investigate the impact of obesity on diastolic function in young subjects.

**Methods:** Prospective echocardiographic measurements were performed in 25 obese children and 25 lean controls. Two-dimensional, M-mode and color M-mode ultrasound, conventional pulse wave Doppler and tissue Doppler imaging (TDI) were used to assess cardiac function.

**Results:** Gender distribution was not different between the two groups but obese children were older (14.1 ± 1 vs 12.9 ± 1.7 yrs,  $p < 0.01$ ) at the time of study. Body mass index (30.4 ± 5.5 vs 18.8 ± 1.7, Z-score 2.58 vs 0.03,  $p < 0.0001$ ), systolic (112 ± 13 vs 101 ± 10 mmHg,  $p < 0.01$ ) and diastolic blood pressure (55 ± 6 vs 52 ± 7 mmHg,  $p < 0.05$ ) were greater whereas heart rate (71 ± 9 vs 77 ± 9 bpm,  $p < 0.05$ ) was lower in the obese group. Measurements of LV mass (132 ± 42 vs 93 ± 25 g,  $p < 0.0001$ ), LV wall thickness (PW 1.28 ± 0.2 vs 1.14 ± 0.17 cm,  $p < 0.01$ ; IVS 1.09 ± 0.2 vs 0.96 ± 0.16 cm,  $p < 0.01$ ), LV end-diastolic diameter (4.8 ± 0.4 vs 4.5 ± 0.4 cm,  $p < 0.01$ )

and volume ( $107 \pm 22$  vs  $91 \pm 22$  ml,  $p < 0.01$ ) were significantly greater in the obese children. Nevertheless, the relative LV thickness  $[(IVS + PW)/LVEDD]$  was not different between the 2 groups. Although LV systolic function (SF and EF) was not different, obese children had a significantly increased systolic ejection volume ( $69 \pm 17$  vs  $55 \pm 16$  ml,  $p < 0.01$ ). LV systolic wall tension was significantly increased in obese children ( $231 \pm 37$  vs  $199 \pm 35$ ,  $p = 0.002$ ) whereas wall stress was not. The mitral E deceleration time ( $131 \pm 24$  vs  $116 \pm 23$  sec,  $p < 0.05$ ), Doppler E/A ratio ( $2.1 \pm 0.5$  vs  $1.8 \pm 0.4$ ,  $p < 0.05$ ), Doppler/TDI E/E' ratio ( $7.1 \pm 1.6$  vs  $6.4 \pm 1.4$ ,  $p < 0.05$ ), TDI E'/A' ratio ( $2.1 \pm 0.5$  vs  $2.4 \pm 0.6$ ,  $p < 0.05$ ) and isovolumic relaxation time ( $65 \pm 12$  vs  $60 \pm 8$  ms,  $p < 0.01$ ) were significantly different between the obese and lean patients, suggesting early LV diastolic dysfunction in obese children.

**Conclusion:** These data confirm that obese children have increased blood pressure and LV mass and suggest an adaptation of the LV dimension to increased LV thickness in order to keep wall stress constant. This leads to increased LV tension and is responsible for early impairment of LV diastolic function. Prevention of obesity is essential in order to prevent irreversible cardiac changes in this young and growing population.

#### P-207

##### **Interest of three-dimensional echocardiography for the assessment of bicuspid aortic valve in children**

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**Introduction:** Bicuspid aortic valve (BAV) is the most common congenital heart defect. Children with BAV are more likely to have valve dysfunction and to require intervention during childhood. According to the subtype of BAV, prognosis and treatment may be different. The aim of this study was to assess the accuracy of 3D echocardiography (3DE) in order to diagnose BAV and to depict accurately the leaflets morphology.

**Methods:** 72 consecutive children with suspicion of BAV were included in a prospective monocentric study. 2DE and 3DE images were analyzed separately by two confirmed pediatric cardiologists in order to assess BAV. We compared 2DE and 3DE for the description of the spatial position of cusps and raphe. The association with aortic aneurysms, aortic coarctation, aortic valve insufficiency or stenosis and other cardiac malformation were also reported.

**Results:** The median age was 6.3 ( $\pm 5.5$ ) years. Using 3DE, BAV was not found in 11.1 [CI 95%, 5.0–20.7] of suspected patients on 2DE. For 22 patients (34.4%) [CI 95%, 22.9–47.3], 3DE allowed a better visualization of the leaflets morphology leading to a reclassification of the BAV. There was a moderate correlation ( $\kappa = 0.57$ ) between 2DE and 3DE for the classification of BAV according to the raphe localization. Inter-observer variability is almost null ( $\kappa = 0.93$ ). Mean time acquisition of 3DE was 2.3 minutes.

**Conclusion:** 3DE is a simple, rapid and reliable method for the diagnosis and the accurate description of BAV in children. This technique may be particularly helpful in order to precise the prognosis or to guide the surgeon.

#### P-208

##### **The role of transesophageal echocardiography in a tertiary congenital heart centre: retrospective study on 1045 consecutive examinations**

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**Introduction:** Transesophageal echocardiography (TEE) has become a major diagnostic tool in care of patients with congenital heart disease in any age group since the development of small pediatric probes. To evaluate the clinical role of TEE in a tertiary congenital heart centre we re-evaluated 1045 consecutive examinations with focus on age, indication, technical procedure and impact.

**Results:** Out of 1045 examinations, 10% were performed in patients less than 1 year of age, 14% in children aged 1 to 6 years, 10% in patients 6–12 years old, 11% in those 12–18 years old, and 55% in adults.

Main indication in the pediatric group was pre- intra and postoperative TEE. (69%), followed by periprocedural TEE at catheter interventions (15%). In the adults, the main indications were catheter interventions (39%), perioperative (23%), and search for thromboembolic sources (18%), suspicion of endocarditis was the indication in 3,2% of pediatric and 4% of adult patients.

A significant impact on intra- and post operative procedure was found in 20%: the rating of regurg after reconstruction of AV-valves made a re-doing necessary, also a persisting shunt after closure of ASD or VSD, an important role is the measuring of the ventricle function while ECMO weaning.

**Conclusion:** Perioperative TEE is the main cause of all TEE in the pediatric group, followed by catheter interventions. In the adult congenital group catheter interventions are the leading indication. Using TEE while catheter interventions the radiation dosage can be reduced. TEE performed in the operation theatre helps to avoid early reoperation because of suboptimal results. All congenital cardiac units should have the TEE option available.

#### P-209

##### **The Impact of Flat-detector computed tomography during Catheterization of congenital Heart Disease**

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**Objectives:** To analyze the diagnostic profit of flat-detector-CT (FD-CT) in the catheterization of patients with congenital heart disease, the help of overlaid 3D-Images on the fluoroscopy and the utility of image-fusion. To develop application protocols for different questions and imaging.



**Methods:** In 2010 we analyzed all our cases where FD-CT was used during catheterization of congenital heart disease. The diagnostic value was determined by the opinion of two pediatric

cardiologists and one surgeon independently. While diagnostic or interventional catheterization, the assistance of image integration in fluoroscopy, either from FD-CT or former MRI or conventional CT (MSCT), was estimated by the operator. The radiation exposure of the FD-CT and contrast volume for rotational angiography was evaluated.

**Results:** 303 cardiac catheterizations were performed in 2010; in 68/303 (20%) we used the new possibility of FD-CT, mainly as angiographic computed tomography (ACT). Median age of the patients was 7.0 years (range 0–42.5). In 35/68 cases the FD-CT-technique was used for diagnostic issue, in 25/68 case we used the 3D-rekonstruktion for navigation in interventions; in 8/68 times we utilized former pictures as MRI-Images or CT-scans for image-fusion in interventions. In over 75% we achieved useful diagnostic images. The median dose-area-product was  $125.1 \mu\text{Gym}^2$  (range 19.3–1295.7), the needed contrast medium was 1.62 ml/kg (range 0–5.7). Applications protocols divers between the varying anatomical regions and the hemodynamic.

**Conclusions:** FD-CT with 3d rotational angiography is a new and very helpful modality, particularly in catheter interventions, also with the option to integrate former pictures from MRI or MSCT and for surgical planning, first of all in the surgery of the great vessels. For the imaging different application protocols are needed. Radiation exposure and needed contrast volume are reasonable.

#### P-210

##### Evaluation of the ventricular functions in the obese children by tissue Doppler echocardiography

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Obese children are predisposed to left ventricular hypertrophy and cardiac dysfunction. The aim of this study was to investigate the impact of childhood obesity on ventricular functions using the conventional and tissue Doppler echocardiography.

Sixty-five obese children were examined by conventional and tissue Doppler echocardiography and compared with 35 lean controls. No differences were evident between lean and obese children in age, height, and gender ( $p > 0.05$ ). Body mass index ( $30.2 \pm 3.6/17.7 \pm 1.8 \text{ kg/m}^2$ ) were significantly greater in the obese children ( $p < 0.001$ ). Systolic and diastolic functions by evaluated conventional echocardiography were normal in obese children. Left ventricle end-diastolic (LVd) diameter, left ventricle-posterior wall end-diastolic (LVPWd) thickness, interventricular septum end-diastolic (IVSd) thickness, left ventricle mass (LVM), left ventricle mass index (LVMI), and relative wall thickness (RWT) were significantly greater in obese children ( $p < 0.001$ ). Body mass index was correlated with the LVPWd, IVSd, LVd, LVM, LVMI and RWT ( $p < 0.001$ ,  $r = 0.78$ ,  $0.76$ ,  $0.61$ ,  $0.79$ ,  $0.75$ , and  $0.48$ , respectively).

Peak late relaxation velocity determined by tissue Doppler echocardiography for the left ventricle ( $A_{dm}$ :  $8.1 \pm 1.8/7.2 \pm 1.4$ ), interventricular septum ( $A_{di}$ :  $7.2 \pm 1.8/6.2 \pm 1.2$ ), and right ventricle ( $A_{dr}$ :  $13.2 \pm 2.6/10.7 \pm 2.5$ ), were significantly higher in obese group ( $p < 0.05$ ). The early to late relaxation velocity ratio for the left ventricle ( $E_{dm}/A_{dm}$ :  $2.4 \pm 0.6/2.9 \pm 0.7$ ), interventricular septum ( $E_{di}/A_{di}$ :  $2.2 \pm 0.7/2.6 \pm 0.8$ ), and right ventricle ( $E_{dr}/A_{dr}$ :  $1.4 \pm 0.2/1.8 \pm 0.5$ ) were significantly lower in obese group compared the control group ( $p < 0.05$ ).  $E/E_{dm}$  was found to be higher in the obese group than the

control group ( $5.9 \pm 1.5/5.1 \pm 1$ ,  $p < 0.01$ ). We found the significantly positive correlation between the LVM and  $A_{dm}$ ,  $E/E_{dm}$  ( $p < 0.05$ ,  $r = 0.24$ ,  $0.22$ , and  $0.22$  respectively). Significantly negative correlation was found between the LVM and  $E_{dm}/A_{dm}$  ( $p < 0.001$ ,  $r = -0.37$ ).

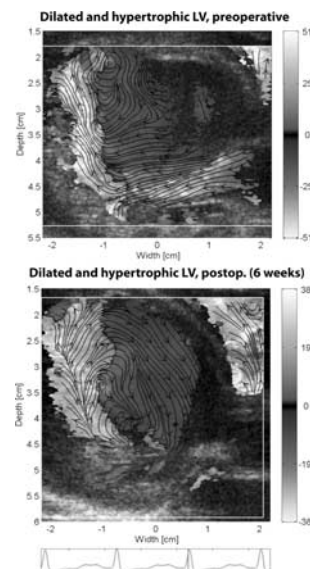
Left ventricular hypertrophy, as evidenced by increased LV mass, was present in obese children. Higher body mass index associated with left ventricle hypertrophy. In our study, tissue Doppler echocardiography revealed subclinical changes in diastolic function of the left ventricle, interventricular septum and right ventricle. These data suggest that indexes of diastolic function, including tissue Doppler measures, was significantly impaired in obese children, and these changes were significantly correlated with LVM.

#### P-211

##### Change in left ventricular vortex flow before and after surgery for coarctation of the aorta in an infant shown with plane wave imaging and flow speckle tracking

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**Introduction:** There has been emerging interest in the relationship between the left ventricular blood flow pattern and left ventricular function. Vortex flow in the left ventricle using magnetic resonance phase-velocity mapping and contrast echocardiography using vector particle image velocimetry has been described, and Doppler ventricular flow pattern may predict left ventricular thrombus after acute myocardial infarction. However, MR is resource demanding, contrast imaging is invasive and old Doppler-techniques are limited in several ways. A new ultrasound flow technique well suited for follow up in children is described. A case with an infant with reduced left ventricular function is presented, where changes in vortex flow before and after surgery for coarctation of the aorta is demonstrated.



**Methods:** We used a high image acquisition rate setup based on transmitting plane wave (unfocused) emissions and by using parallel receive processing to generate several image lines simultaneously. Using pattern-matching techniques, the movement of blood speckle was quantified in 2-D images. The 2-D velocity information can be visualized as arrows or streamlines



overlaid colour-Doppler images that help to highlight areas of complex flow patterns such as vortex formation. Using plane wave imaging we are currently limited to using linear array transducers (here we used a 9L linear transducer). As the need for angle-correction is removed an increased accuracy in quantitative measurements may also be obtained.

**Results:** The figures demonstrate the left ventricular flow pattern in a 5.5 months old girl with coarctation of the aorta and left ventricular dysfunction. The upper image demonstrates the left ventricular vortex flow pattern at admission. Please observe apical vortex ring formation. The lower image demonstrates the left ventricular flow pattern six weeks after surgical repair when left ventricular function has improved, but the ventricle still was dilated with reduced contractility. Here the vortex is in the central part of the ventricle and there is improved emptying of the left ventricle.

**Conclusions:** This case report demonstrates that abnormal left ventricular function may be reflected in the left ventricular flow patterns which we can accurately describe using this non invasive ultrasound flow tracking technique.

#### P-212

##### **Echocardiographic evaluation of pulmonary vascular resistance and reactivity in children with pulmonary hypertension**

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**Introduction and Aim:** Pulmonary vascular resistance and vasoreactivity are the most important prognostic factors in childhood pulmonary hypertension. The present study aims to determine the power and effectivity of transthoracic Doppler echocardiography in diagnosis of pulmonary vascular resistance and vasoreactivity by performing transthoracic echocardiography and cardiac catheterization simultaneously.

**Materials and Methods:** The present study reviews 40 healthy controls and 30 children with pulmonary hypertension as diagnosed by cardiac catheterization. For each subject, transthoracic Doppler echocardiography was done simultaneously during the period cardiac catheterization was performed. When examination by transthoracic echocardiography was carried out at the distal region of the right ventricle outflow tract, Doppler determining indices including acceleration time (AcT), inflection time (InT), deceleration index (DI), deceleration time (DecT), right ventricle ejection time (RVET), AcT/RVET, TRPG/RVOTVTI, Ts/RVOTVTI, PAB/HRRVOT and their values adjusted for heart beat rate were recorded. Vasoreactivity test by O<sub>2</sub> was done in children with pulmonary hypertension so that any alteration in the parameters could be specified.

**Results:** The mean age of the children with pulmonary hypertension was  $77.6 \pm 93.2$  months while the mean age of the healthy control subjects was  $77.6 \pm 64.8$  months. The AcT, AcTc, InT, InTc, DI, DIc, DecT, DecTc, AcT/RVET parameters of the children with pulmonary hypertension were statistically different from those of the healthy control subjects ( $p < 0.001$ ;  $p < 0.001$ ;  $p < 0.001$ ;  $p < 0.001$ ;  $p = 0.02$ ;  $p = 0.01$ ;  $p < 0.001$ ). Children with pulmonary hypertension were grouped into three according to their pulmonary vascular resistance (namely 0–3, 3–10 and >10). It was also found that AcTc, TRPG/RVOTVTI, Ts/RVOTVTI, and PAB/HRRVOT were indicators of an increase in pulmonary vascular resistance ( $p = 0.009$ ;  $p = 0.009$ ;  $p = 0.003$ ;  $p = 0.004$ ). Moreover there

was a statistically significant difference in the parameters of AcTc, DIc, DecT, TRPG/RVOTVTI, Ts/RVOTVTI, and PAB/HRRVOT of the patients who responded to the vasoreactivity test ( $p = 0.005$ ;  $p = 0.009$ ;  $p = 0.014$ ;  $p = 0.004$ ;  $p = 0.002$ ;  $p < 0.001$ ).

**Discussion:** Pulmonary vascular resistance and vasoreactivity can be evaluated by the examination of pulmonary artery flow-velocity curves with the help of transthoracic Doppler echocardiography. Transthoracic Doppler echocardiography seems to be an effective test for screening increased pulmonary vascular resistance and vasoreactivity in children with pulmonary hypertension.

#### P-213

##### **Role of speckle strain for evaluation of ventricular function after Total Cavo-Pulmonary Connection: does it identify patients at risk?**

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**Introduction:** The evaluation of ventricular function of patients with total cavopulmonary connection (TCPC) is the object of discussion. In fact, the parameters used to evaluate systolic and diastolic function of left and right ventricle can not be applied to single ventricles with large anatomical variability. We investigated the role of speckle strain echocardiography in this setting.

**Patients and methods:** From January 1995 to December 2010, 68 patients had TCPC; 58 are regularly followed. Among these we evaluated 38 patients aging >10 years (median age 14 years, median distance from TCPC 7 years) with trans-thoracic M-mode, 2D, Doppler, Color-Doppler and systolic strain rate (18 segments). No patient had overt ventricular dysfunction; 2/3 of patients (group A) had a normal life including normal physical activity, normal oxygen saturation and had never needed interventional catheterization or reinterventions. The remaining patients had cyanosis, limited physical capacity and had needed repeated interventions (group B). In this group 2 patients had plastic bronchitis and 1 protein losing enteropathy.

**Results:** Mean fractional shortening was  $29 + 8.6\%$ , mean biplanar EF was  $53 + 4.5\%$  and similar in both groups ( $53.9 + 4.1$  versus  $51.6 + 5.6$ ). Mean ventricular mass was  $112.7 + 31.7 \text{ g/m}^2$  and significantly higher in group A patients ( $122.9 + 23.8$  versus  $84.8 + 36.9$ ,  $p < 0.03$ ). Mean systolic strain rate was  $-16.1 + 2.2$  and similar in both groups ( $-16 + 2$  versus  $-16.6 + 2.8$ ). EF and strain rate were significantly related ( $p < 0.01$ ).

**Conclusions:** Our data show that ventricular function was generally normal either in patients with excellent or poor clinical condition. In particular, systolic strain rate was normal or mildly depressed in the global population, while a higher ventricular mass might identify patients in good clinical condition. Echographic evaluation of systolic function in itself, in the absence of overt ventricular dysfunction, does not prove to be a “marker of health” in this population.

#### P-214

##### **Right ventricular volumes and function assessed by three-dimensional echocardiography in children with surgically corrected Tetralogy of Fallot and their healthy controls: Comparison to cardiac MRI**

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**Objectives:** Deterioration of right ventricular (RV) function is common in patients with tetralogy of Fallot (TOF) because of chronic volume loading due to pulmonary regurgitation. Severe dysfunction and dilatation might be irreversible and therefore regular assessment of RV function is indicated. Cardiac magnetic resonance imaging (CMR) is still the golden standard for these measurements. A more available, less expensive and faster method is real time 3-dimensional echocardiography (RT3DE), which is already proven to be adequate in adult patients with TOF. The aim of this study was to determine if the 3DE could be used for assessment of RV function already in children and adolescents.

**Methods:** 50 pediatric patients (mean age  $13.1 \pm 3.1$  years,) who had undergone TOF repair and their 45 healthy age and gender matched controls underwent RT3DE-study to assess the RV ejection fraction (EF), end diastolic volume and end systolic volume (EDV and ESV). CMR-study was performed as a reference.

**Results:** RT3DE data quality was sufficient in 79% of all subjects examined. When RT3DE was compared with CMR EDV was  $97 \pm 27 \text{ ml/m}^2$  vs.  $100 \pm 28 \text{ ml/m}^2$  ( $p = 0.42$ ), ESV was  $46 \pm 16 \text{ ml/m}^2$  vs.  $48 \pm 20 \text{ ml/m}^2$   $p = 0.50$  and EF  $55 \pm 8.4\%$  vs.  $51 \pm 6.6\%$ , respectively ( $p = 0.0023$ ). Good correlation between these two methods was shown in all measurements, EDV ( $r = 0.51$ ,  $p < 0.0001$ ), ESV ( $r = 0.44$ ,  $p = 0.006$ ) and EF ( $r = 0.38$ ,  $p = 0.04$ ).

**Conclusions:** CMR is the standard method for measuring the RV function but is time consuming and expensive. Our study shows that RT3DE is a valuable, faster tool in assessing RV volumes and EF already in pediatric patients. RV volumes measured with RT3DE -study correlate fairly well with CMR. We believe that RT3DE is a promising tool in assessment of RV volume and function in ambulatory patients also in children.

#### P-215

##### **Visualization of the Intracavitary Blood Flow in Systemic Ventricles of Fontan Patients by Contrast Echocardiography Using Vector Particle Velocimetry.**

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**Introduction:** Diastolic vortex formation in Fontan patients may have an impact on cardiac function. Therefore, this study aimed at assessing and characterizing vortex flow patterns in patients with Fontan circulation in comparison with healthy controls.

**Methods:** Twenty-six patients (11 fontan and 15 normal patients) underwent echocardiography with intravenous contrast agent (Sonovue®) administration. Dedicated software was used to perform particle image velocimetry (PIV) and to visualize intracavitary flow in the systemic ventricles of the patients. Vortex parameters including vortex depth, length, width, and sphericity index were measured. Vortex pulsatility parameters including relative strength, vortex relative strength, and vortex pulsation correlation were also measured.

**Results:** Vortex length (VL) was significantly lower in Fontan patients ( $0.543 \pm 0.102$  vs  $0.651 \pm 0.125$ ,  $p = 0.024$ ). Vortex width (VW) was higher ( $0.363 \pm 0.093$  vs  $0.276 \pm 0.044$ ,  $p = 0.014$ ) and sphericity index (SI) was lower ( $1.566 \pm 0.439$  vs  $2.421 \pm 0.626$ ,  $p = 0.001$ ) in the normal group. Relative strength (RS) ( $0.756 \pm 0.517$  vs  $1.903 \pm 0.471$ ,  $p < 0.0001$ ) and vortex relative strength (VRS) ( $0.190 \pm 0.113$  vs  $0.433 \pm 0.141$ ,  $p < 0.0001$ ), were significantly lower in the Fontan patients group.

**Conclusions:** Fontanpatients had aberrant flow patterns as compared to normal hearts in terms of position, shape, sphericity and direction of the main vortices. Whether vortex characteristics are related with clinical outcome is subject to further investigation.

#### P-216

##### **The functional right ventricle in patients with Ebstein's anomaly is not small**

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**Introduction:** Contemporary opinion regarding the size of the functional right ventricle (RV) in patients with Ebstein's anomaly is that the functional RV is small. However, only few have attempted to measure the size of the functional RV in post-mortem hearts of patients with Ebstein's anomaly. Therefore, in vivo size of the functional RV in patients with Ebstein's anomaly is actually still unknown. Therefore, the aim of this study was to determine the size of the functional RV of patients with Ebstein's anomaly without previous cardiac surgery by cardiovascular magnetic resonance.

**Methods:** Cardiovascular magnetic resonance of 36 consecutive patients with unoperated Ebstein's anomaly (mean age  $33 \pm 20$  yrs, 23 female, February 2006 till December 2010) determined functional RV volume. Ventricular volumes were measured using standard analysis software.

Additionally, in a subgroup of 11 patients with Ebstein's anomaly short axis and axial slices were performed to measure ventricular volumes and to determine inter- and intra-observer variances.

**Results:** Mean Right Ventricular Enddiastolic Volume (RVEDV) was  $256 \pm 141 \text{ ml}$ ; RVEDV indexed to body surface area (RVEDVI) was  $154 \pm 70 \text{ ml/m}^2$ . Maximum values were  $656 \text{ ml}$  and  $365 \text{ ml/m}^2$ , respectively. Only 4 Patients had normal RVEDVI  $< 90 \text{ ml/m}^2$ . RVEDV: LVEDV ratio was  $2.9 \pm 1.7:1$ . Maximum RVEDV:LVEDV was  $8.8:1$ .

Inter- and intra-observer variance were not significantly different between axial and short axis slice measurements.

**Conclusions:** Our results show that the volume of the functional RV in patients with Ebstein's anomaly is not small. Only 4 patients had normal RVEDVI. None of the patients had a small functional RV. Therefore, contemporary opinion regarding the size of the functional RV in patients with Ebstein's anomaly should be revised.

#### P-217

##### **Pulmonary and systemic vascular resistance in patients with open and closed ASD type secundum.**

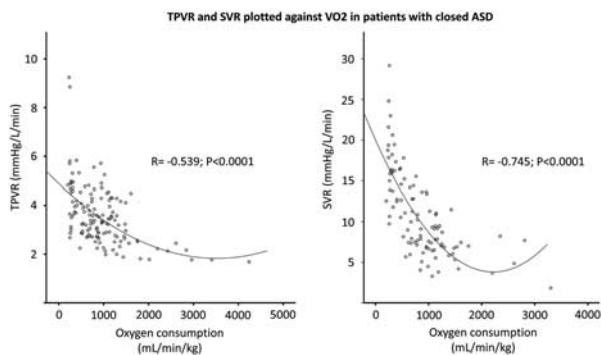
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**Introduction:** This study aimed at (1) evaluating total pulmonary vascular resistance (TPVR) and systemic vascular resistance (SVR), (2) comparing the relative change in TPVR and SVR and (3) evaluating the relationship between oxygen consumption and TPVR/SVR during exercise in patients with open and closed ASD type secundum.

**Methods:** Fourteen patients with open (mean age  $39 \pm 19$  years) and 29 patients with closed (mean age  $41 \pm 17$  years) ASD

underwent standard and symptom-limited bicycle stress echocardiography. Oxygen consumption (VO<sub>2</sub>) was measured using cardiopulmonary exercise testing. TPVR was calculated as the ratio of systolic pulmonary artery pressure to right ventricular cardiac output and SVR was calculated as the ratio of mean blood pressure to left ventricular cardiac output at each stage.

**Results:** TPVR decreased significantly from rest to peak exercise in patients with closed ASD. ( $4.6 \pm 1.6$  to  $3.3 \pm 0.9$  mmHg/L/min;  $P < 0.0001$ ) but not in patients with open ASD. ( $3.1 \pm 0.9$  to  $3.1 \pm 0.7$ ;  $P = 0.940$ ). SVR decreased significantly from rest to peak exercise in patients with a closed ( $22.3 \pm 4.7$  to  $11.5 \pm 2.7$  mmHg/L/min) and open ASD. ( $24.8 \pm 5.9$  to  $13.4 \pm 5.8$  mmHg/L/min). The decrease in SVR was larger than the decrease in TPVR in patients with open ( $-47 \pm 27$  versus  $+5 \pm 33\%$ ;  $P < 0.0001$ ) and closed ( $-47 \pm 15$  versus  $-19 \pm 26\%$ ;  $P < 0.0001$ ) ASD. In patients with a closed ASD, there was an inverse relation between TPVR and VO<sub>2</sub> ( $R = -0.539$ ,  $P < 0.0001$ ) and between SVR and VO<sub>2</sub> ( $R = -0.745$ ,  $P < 0.0001$ ). In patients with an open ASD, there was an inverse relation between SVR and VO<sub>2</sub> ( $R = -0.773$ ,  $P < 0.0001$ ). These relationships were best fit by a quadratic equation (figure).



**Conclusions:** TPVR does not decrease during exercise in patients with an open ASD. SVR decreases significantly and similarly in patients with an open and closed ASD. As expected, the relative change in SVR was larger than the relative change in TPVR. There was an inverse relation between total PVR and VO<sub>2</sub> in patients with closed ASD, which could best be described by a quadratic equation.

#### P-218

##### The Coronary Arteries in Patients with Hypoplastic Left Heart Syndrome – An Angiographic Study and its Clinical Implications

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**Introduction:** Coronary anatomy in hypoplastic left heart syndrome (HLHS) has attracted increasing attention. This is the first angiographic study of coronary artery anatomy in HLHS while prior descriptions mostly based on post-mortem examinations.

**Methods:** Coronary angiograms obtained by selective native aortic root injection were reviewed in 84 patients (mitral atresia/aortic atresia,  $n = 39$ ; mitral stenosis/aortic stenosis,  $n = 25$ ; mitral stenosis/aortic atresia,  $n = 13$ ; mitral atresia/aortic stenosis,  $n = 7$ ). Origin and epicardial course, anomalies, coronary dominance and native ascending aorta dimensions were analyzed.

**Results:** Right dominance was present in 43 (51.2%), left in 31 (36.9%) and balanced type in 10 (11.9%) patients. Coronary dominance was unrelated to anatomic subtypes ( $p = 0.163$ ), but left dominance and balanced type was more common in patients with absent left ventricular cavity ( $p = 0.011$ ). Coronary artery fistulas were found in 15 (17.9%) and tortuosity of the coronary arteries, mostly affecting the left anterior descending artery, in 28 (33.3%) patients. Both occurred more frequently with mitral stenosis/aortic atresia (Tortuosity 12 of 13 patients,  $p \leq 0.001$ ; Coronary artery fistulas 6 of 13 patients,  $p = 0.001$ ). Collaterals to extra-cardiac vessels were visualized in 41 patients. The native ascending aorta was smaller with aortic atresia ( $44.4 \pm 20.5$  mm<sup>2</sup>/m<sup>2</sup> vs.  $127.8 \pm 71.8$  mm<sup>2</sup>/m<sup>2</sup>,  $p \leq 0.001$ ). In 18 (21.4%) patients with relatively large native ascending aorta ( $113.8 \pm 69.2$  mm<sup>2</sup>/m<sup>2</sup> vs.  $65.9 \pm 56.2$  mm<sup>2</sup>/m<sup>2</sup>) retention of contrast media in the aortic root identified areas of low blood flow. One of these patients experienced myocardial infarct and cerebral stroke 7 years after catheterization and thrombus formation was detected within the aortic root.

**Conclusions:** Left dominance was more prevalent compared to the normal population. Observed anomalies were tortuosity, coronary artery fistulas and collateral vessels. The latter could be interpreted as a result of repeated surgical interventions. The long-term effect of these findings is unclear. Most coronary artery fistulas are small and coronary perfusion seems not to be altered. Native ascending aorta dimension remained larger in patients with aortic stenosis. This might bear a risk for thrombus formation in the aortic root. Anticoagulation might be warrantable.

#### P-219

##### Changes in Myocardial Velocities in First week of life in Pre Term Infants.

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**Introduction:** Significant haemodynamic changes occur during fetal-neonatal transition: the patent ductus arteriosus closes, pulmonary blood flow increases, preload and afterload of the Left ventricle increases, right ventricle afterload decreases, and the contractility and structure of myocardium change, all of which may impact on diastolic function. To date no validated technique allows accurate assessment of diastolic function in this population. Tissue Doppler imaging (TDI) may be a novel technique to measure diastolic function over the first week of life in the preterm infant.

**Aims:** To use tissue Doppler imaging as a modality to assess diastolic function in preterm infants during the first week of life.

**Methods:** Preterm infants less than 32 weeks gestation or <1500 g born in the National Maternity Hospital were recruited and ethical approval and written consent were obtained. Echocardiography was carried out by a single observer using the GE Vivid I machine, on Day 1, 3–4 and Day 7. Myocardial velocities were obtained using a pulsed wave doppler sample from the lateral mitral and tricuspid annuli from an apical four chamber view. Peak systolic (S'), early diastolic (E') and late diastolic (D') velocities were recorded.

**Result:** Twelve infants with structurally normal hearts were recruited to date. Gestational age range was 24+4–31+5 weeks and heart rate varied from 149–168 bpm in the first week.

There was an increase in myocardial velocities across all measurements in the first week of life and in all infants all infants peak velocities were higher in the right ventricle than the left.

Table 1: Variations in myocardial velocities in the first week of life in preterm infants.

Preterm Infants	Right Ventricle			Intra ventricular Septum		
	S'	E'	A'	S'	E'	A'
D1	3.96	3.69	7.35	2.75	3.74	4.85
D3-4	4.04	4.3	7.6	2.88	3.37	4.1
D7	4.9	5.5	9.1	2.1	3.04	5.25

Preterm Infants	Left Ventricle		
	S'	E'	A'
	3.5	3.99	5.18
	3.28	4.16	5.58
	3.41	4.41	5.4

**Conclusions:** In this small cohort TDI is a feasible measure of diastolic function in the preterm infant.

#### P-220

##### **Detection of right ventricular fibrosis by CMR and plasma levels of procollagen type III N-terminal amino peptide in patients with Tetralogy of Fallot**

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**Objective:** Recently the serum concentration of aminoterminal procollagen type III (PIIIP) was shown to be elevated in patients with congenital heart disease (CHD). Ventricular myocardial fibrosis is also evident in the right ventricle of patients after correction of Tetralogy of Fallot (TOF). The levels of PIIIP are associated with the severity of haemodynamic load or hypoxaemia, which both might induce myocardial fibrosis. We hypothesized that serum levels of PIIIP would correlate with the amount of fibrosis detected by late gadolinium enhancement (LGE) cardiovascular magnetic resonance (CMR). We also hypothesized that the amount of fibrosis would show correlation with other clinical markers such as pro-BNP and the right ventricular volume.

**Methods:** Serum PIIIP levels were measured in two groups: in 50 pediatric patients (mean age 13.1 years, SD 3.1) who had undergone TOF repair and in 43 healthy age and gender matched controls. These groups underwent CMR-study and LGE was scored in the right and left ventricle (RV and LV).

**Results:** PIIIP levels of TOF patients were significantly higher than those of control subjects (12.4 SD 4.9 vs. 8.3 SD 2.9  $p < 0.0001$ ). RV LGE was found in all of the patients but LGE score did not correlate with PIIIP levels ( $p = 0.16$ ). LGE score correlated positively with the RV end diastolic volume (EDV ml/m<sup>2</sup>  $p < 0.016$ ) and with pro-BNP ( $p < 0.015$ ). No LV LGE was shown in any of the patients and both LGE scores were zero in all control subjects. PIIIP levels did not correlate with EDV or pro-BNP.

**Conclusions:** RV LGE is a common finding in TOF patients already in pediatric age whereas LV LGE is not present at all. Cardiac collagen turnover is known to be active in myocardium under pathological conditions. We suggest that the elevated levels

of PIIIP reflect an active turnover process in the RV, but the amount fibrosis can not be estimated by plasma PIIIP levels. Higher LGE score is associated with more dilated right ventricle and suggests that dilatation rather than restriction is common in the right ventricle of TOF patients.

#### P-221

##### **Correlation of the right atrial volume with the right ventricular end-diastolic pressure**

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**Introduction:** The right ventricular end-diastolic pressure (RVEDP) is an important parameter for the assessment of the right ventricular function. It is determined invasively by cardiac catheterization. The right atrium is directly subjected to the RVEDP through the open tricuspid valve. In case of elevated pressure the right atrium is susceptible to dilation due to its thin-walled structure. This suggests that the right atrial volume evaluated non-invasively by cardiovascular magnetic resonance imaging (CMR) can indicate chronic diastolic dysfunction of the right ventricle.

**Methods:** 35 patients with dilated right ventricle due to chronic pulmonary regurgitation were subjected to cardiac catheterization and CMR to evaluate the necessity of pulmonary valve replacement. In CMR the regurgitant fraction of the pulmonary artery and the end-diastolic right ventricular volume were determined. In addition, the right atrial volume was assessed using ECG-gated cine steady state free precession (SSFP) sequences for the acquisition of gapless slices in the axial plane: In each slice the endocardium was manually contoured, and the resulting volumes were added according to the modified Simpson's rule. Patients with tricuspid insufficiency were excluded from the study.

**Results:** The normalization of the right atrial volume to body surface area resulted in the right atrial volume index (RAVI), which was on average  $60.8 \pm 20.1$  ml/m<sup>2</sup>. Cardiac catheterization revealed a mean RVEDP of  $10.4 \pm 2.7$  mmHg, each of the values correlating well with the respective right atrial pressure (mean  $9.1 \pm 2.6$  mmHg,  $r = 0.91$ ,  $p < 0.001$ ). Furthermore, the correlation between RVEDP and RAVI was shown to be statistically significant ( $r = 0.45$ ,  $p = 0.007$ ) despite scatter of individual values.

**Conclusions:** The right atrial volume index determined by cardiovascular magnetic resonance imaging correlates well with RVEDP. Hence it is a parameter for diastolic dysfunction of the right ventricle that can be assessed non-invasively.

#### P-222

##### **Exercise testing coupled with Doppler echocardiography: a simple, safe and effective method to assess the severity of coarctation of the aorta**

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Peak systolic Doppler velocity at the aortic isthmus alone is not a good predictor of severity in patients with native or residual coarctation after surgical repair. Evaluating the need for surgery or percutaneous stenting is often difficult in these cases. A major argument in favour of a hemodynamically significant aortic

isthmus narrowing is the presence of a diastolic gradient with characteristic “sawtooth” appearance of the Doppler pattern.

**Objectives:** to evaluate, in patients with suspected aortic isthmus narrowing but without any significant diastolic gradient at rest, if exercise could unmask diastolic gradient and thus reveal a hemodynamically significant coarctation.

**Methods:** Fourteen patients aged from 12 to 56 years underwent treadmill exercise testing coupled with Doppler echocardiography. Thirteen had previous coarctation repair 8 to 43 years before, one had aortic kinking with mild diffuse isthmus narrowing on CT-scan. MRI or CT-scan were performed in 11 patients, significant residual stenosis (narrowing >30%) was detected in 8/11. Exercise testing was maximal or juxtamaximal in all but 1. Doppler measurements were performed during exercise testing and 5 minutes after at the suprasternal notch, using a CW Doppler 2MHz pedoff probe. The peak systolic and diastolic gradients through aortic isthmus were measured at rest and at the end of the exercise. A peak protodiastolic gradient > 17 mmHg was considered as significant, as described in previous studies.

**Results:** Mean peak systolic gradient increased from 29 to 65 mmHg with exercise (mean increase +142%,  $p < 0.001$ ). Among the eight patients without any diastolic gradient at rest, significant diastolic gradient appeared in 4. Among the 6 patients with non significant diastolic gradient at rest, the gradient remained non significant in 2 and became significant in 4.

**Conclusion:** Apparition of a significant isthmus Doppler diastolic gradient at exercise can reliably predict the hemodynamic significance of aortic restenosis after coarctation repair, as well as native narrowing of the aorta. This simple, safe and effective non-invasive method may be helpful to identify patients requiring surgery or percutaneous stenting. It could also be useful for the follow-up of patients with a coarctation operated on in infancy.

#### P-223

##### **New magnetic resonance imaging indexes for the evaluation of elastic properties of aortic wall in patients with bicuspid aortic valve**

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**Introduction:** Bicuspid aortic valve (BAV) is frequently associated to aortic wall abnormalities as dilation of ascending aorta (AAo) and even dissection.

**Aim:** to compare BAV patients with matched control subjects, prospectively evaluating two new indexes of aortic wall biophysical properties: maximal rate of systolic distension and diastolic recoil (MRSD and MRDR respectively).

**Methods:** we enrolled 53 consecutive young patients with BAV. (36 male, mean age  $16 \pm 4$  years), with no more than mild aortic valve disease, and 22 age- and sex-matched healthy volunteers as control group. All subjects underwent a cardiac MRI study comprehensive of phase velocity mapping acquisition at several aortic level. We defined two new indexes of aortic wall biophysical properties, briefly the cross sectional area of ascending aorta (5 mm over the sino-tubular junction) was measured in each cardiac phase and expressed as a percentage of the maximal end systolic cross-sectional area. The variation in

percentile of maximal area was measured by the difference between each cardiac phase and the previous one (percentile of maximal area/10–3 sec). Thus, MRSD was defined as the maximal systolic increase of ascending aorta cross-sectional area in percentile and MRDR as the highest reduction in percentile of cross sectional area during diastolic recoil MRSD and MRDR were measured in the ascending aorta in patients with BAV and in controls.

**Results:** 26 patient with BAV had enlarged AAo (dilated BAV), 27 had normal diameters (non-dilated BAV). Compared to controls, MRSD was significantly lower in the whole BAV group ( $4.37 \pm 1.1$  vs  $9.1 \pm 2.1$ ), in dilated-BAV. ( $4.5 \pm 1.1$   $p < 0.0001$ ), and non-non dilated BAV. ( $4.3 \pm 1.0$ ,  $p < 0.0001$ ). MRDR was higher in the whole BAV group ( $-4 \pm 1.2$  vs  $-7.6 \pm 2.7$ ,  $p < 0.0001$ ), in dilated BAV ( $-3.9 \pm 1.3$ ,  $p < 0.0001$ ) and in non-dilated BAV ( $-4.1 \pm 1.2$ ,  $p < 0.0001$ ). At ROC curve analysis MRSD was able to distinguish BAV from controls with a sensibility of 100% and specificity of 95%.

**Conclusion:** MRSD and MRDR were slower in BAV patients than in controls independently of the dimension of ascending aorta.

#### P-224

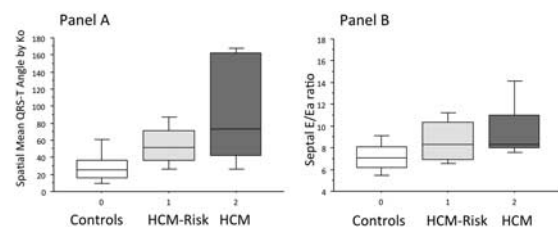
##### **Tissue Doppler Imaging combined with advanced 12-lead ECG analysis might improve early diagnosis of hypertrophic cardiomyopathy in childhood**

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**Introduction:** Optimization of early diagnosis of childhood hypertrophic cardiomyopathy (HCM) is essential in lowering the risk of HCM complications. Standard echocardiography (ECHO) has shown to be less sensitive in this regard. In this study, we sought to assess whether spatial QRS-T angle deviation, which has shown to predict HCM in adults with high sensitivity, and myocardial Tissue Doppler Imaging (TDI) could be additional tools in early diagnosis of HCM in childhood.

**Methods:** Children and adolescents with familial HCM ( $n = 10$ , median age 16, range 5–27 years), and without obvious hypertrophy but with heredity for HCM ( $n = 12$ , median age 16, range 4–25 years, HCM or sudden death with autopsy-verified HCM in  $\geq 1$  first-degree relative, HCM-risk) were additionally investigated with TDI and advanced 12-lead ECG analysis using Cardiax<sup>®</sup> (IMED Co Ltd, Budapest, Hungary and Houston). Spatial QRS-T angle (SA) was derived from Kors regression-related transformation. Healthy age-matched controls ( $n = 21$ ) were also studied. All participants underwent thorough clinical examination.



**Results:** Spatial QRS-T angle (Figure/Panel A) and septal E/Ea ratio (Figure/Panel B) were most increased in HCM group as compared to the HCM-risk and control groups ( $p < 0.05$ ). Of note, these 2 variables showed a trend toward higher levels in HCM-risk group than in control group ( $p = 0.05$  for E/Ea and 0.06 for QRS/T by ANOVA). In a logistic regression model, increased SA and septal E/Ea ratio appeared to significantly



predict both the disease (Chi-square in HCM group: 9 and 5, respectively,  $p < 0.05$  for both) and the risk for HCM (Chi-square in HCM-risk group: 5 and 4 respectively,  $p < 0.05$  for both), with further increased predictability level when these 2 variables were combined (Chi-square 10 in HCM group, and 7 in HCM-risk group,  $p < 0.01$  for both).

**Conclusions:** In this small material, Tissue Doppler Imaging and spatial mean QRS-T angle deviation, particularly when combined, appear to be sensitive in predicting the risk for developing childhood HCM. Large-scale, prospective studies are needed to confirm this hypothesis.

## P-225

### 2D Speckle Tracking assessment of left ventricular torsion in healthy children and heart transplants

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**Introduction:** During cardiac cycle the left ventricular (LV) base and apex rotate in opposite directions resulting in a twisting motion. These movements can be assessed by 2D Speckle tracking echocardiography (2DSTE), a new, non-invasive echocardiographic method. Our aim is to study the feasibility and the reproducibility of LV rotation parameters in healthy children (HC) by 2DSTE, describe normal parameters of rotation in HC and compare them with children undergoing heart transplantation (HT).

**Methods:** 30 HC and 40 HT were included prospectively, all in sinus rhythm, no congenital heart disease nor acute rejection. An iE33 Philips ultrasound system with QLAB software was used. Two observers (X and Y) completed all the measurements: systolic global and endocardial peak rotation at the base and the apex. Torsion was calculated as the difference between base and apex peak rotation.

**Results:** No significant differences were found among both groups, HC and HT, in relation with gender (67,7% vs 54,8% male), mean age (7,7 vs 9,8 years), mean weight (29,1 vs 32,1 kg) or body surface area (0,98 vs 1,05m<sup>2</sup>). In the HC group mean global and endocardial torsion by observer X were 6,2° and 8,4°; by observer Y 6,1° and 7,8°. For global torsion the Lin's concordance correlation coefficient of absolute agreement was 0,72 and for endocardial torsion 0,74. The Bland-Altman interval of agreement between the two observers mean global measures was -0,13° (95% Agreement Interval: -3,40 to 3,13) and the mean endocardial measures was -0,71° (95% Agreement Interval: -5,48 to 4,06). Mean global torsion is slightly higher but not significantly different in patients undergoing HT than in HC. (7,2° vs 6,1°  $p = 0,1$ ), neither is mean endocardial torsion (9,2° vs 8,1°  $p = 0,27$ ).

**Conclusions:** Measures of LV rotation and torsion by 2DSTE in healthy and heart transplant children is feasible and reproducible with a good inter-observer agreement. Although global and endocardial torsion are higher in heart transplants, no statistically significant differences among both groups.

## P-226

### Coronary arteries originating from the opposite coronary cusp: diagnosis and long term outcome

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Coronary arteries originating from the opposite coronary cusp with inter arterial coursing may cause sudden death in children and young adults during exercise.

This study was to review the clinical and echoDoppler data of patients diagnosed with isolated coronary arteries originating from the opposite coronary cusp.

**Material and Methods:** Functional symptoms, coronary anatomy, treatment and survival were reviewed.

**Results:** 28 patients, 17 males and 11 females, were included in the study. Age at diagnosis was 3 days to 76.2 years (mean 21.2y, median 11y) : Seven had no symptom (including 1 neonate and 1 heart transplanted patient), 21 experienced symptoms at exercise: resuscitated sudden death (1), myocardial necrosis (1), syncopes (2) chest pain episodes (7), chest pain and lipothymia (5), near-miss (2), pulmonary edema (2), tachycardia (2).

All patients had careful echoDoppler study and colour Doppler analysis was critical to assess coronary artery blood flow and detect interarterial course (first diagnosis was made prior to 2Decho, either by CTscan in 5 cases or angiography in 3): 12 had RCA arising from left coronary sinus, 10 LCA from right coronary sinus, 6 single coronary arteries from R cusp including 2 RV infundibular coursing and 2 coursing anteriorly to PA. Thirteen patients (46%) underwent surgical reimplantation of the coronary artery (12cases), at the mean age of 14.5 years (1month to 55 years, median 20 years) or coronary stenting (RCA: 1case). Fifteen were not operated on because of young age (1), transplanted heart (1), no inter arterial coursing (6) incidental diagnosis (4) or patient refusal (3). One newborn operated at one month of age because of biventricular dysfunction died 3 month later of cardiac failure despite uneventful reposition of an interarterial RCA. Autopsy showed associated non ischemic cardiomyopathy. The others 12 operated patients had no recurrent symptoms at a mean follow up of 7 years (1to18 years) Non operated patients are still asymptomatic.(mean FU 3,8 years)

**Conclusion:** EchoDoppler can detect coronary arteries originating from the opposite coronary cusp with inter arterial coursing and allow prompt surgical management in symptomatic patients with favourable outcomes. Surgical treatment remains matter of debate in asymptomatic cases with incidental diagnosis.

## P-227

### Dobutamine stress echocardiography in the assessment of postoperative left ventricular function in children after arterial switch operation for transposition of great arteries

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**Background:** Dobutamine stress echocardiography (DSE) is an established indirect method of assessing coronary circulation. It is used to diagnose coronary artery disease and coronary circulatory disorders in the course of congenital and acquired pathology.

**Aim:** The aim of this study was to evaluate the usefulness of DSE in the assessment of postoperative left ventricular function in patients after arterial switch operation (ASO) for transposition of great arteries (TGA).

**Material and Methods:** The study group consisted of 70 patients (51 boys, 25 girls) who underwent neonatal arterial switch operation. 53 pts (70%) had TGA with an intact ventricular septum, 17 (22%) – TGA with ventricular septal defect, 6 – with the pathology of the aortic arch. 34 children (48%) had unusual coronary patterns or coursing. Patients were assessed at the age of 3 to 16 years (mean 7.9 ± 2.69). All patients underwent DSE according to the established protocol. Dobutamine was infused in

3-minutes stages with doses of 5 to 40mcg/kg/min and atropin at 0.01 mg/kg when needed. Echocardiographic images were obtained in 4 views using 17-segmental model. A positive test response was defined as a new or worsened wall motion abnormalities. All patients underwent at least one selective coronary angiography.

**Results:** 53 (69.7%) of 76 studies were normal, 7 were non diagnostic (in 5pts the test was interrupted). In 16 pts DSE was positive (4 with abnormal coronary angiography) without clinical symptoms. All studies were performed without major complications. Adverse events occurred in 25 children (headache, abdominal pain, arrhythmia). None of the patients required treatment and resolved after discontinuation of drug infusion.

**Conclusions:** DSE is an useful, safe and capable of repeating method of postoperative evaluation of TGA patients after arterial switch operation. It may be helpful in targeting patients with increased risk of coronary events in the long-term postoperative period after arterial switch operation.

#### P-228

##### RV-dysfunction – Does it affect left ventricular torsion?

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**Background:** Right ventricular dysfunction can induce changes in left ventricular deformation by RV/LV interaction. We investigated the influence of changes in RV-loading conditions as well as RV-Pacing on LV-rotation.

**Methods:** 20 patients with ASD (median 8 y), 18 patients with valvular pulmonary stenosis (PST, median 2.9 y) and 50 patients with chronic RV-pacing (PM, median 14 y) were examined with a Vivid 7 ultrasonic device. Patients undergoing interventions were examined 4 hours after the procedure again. Healthy children served as body-surface area matched controls. Maximal torsion and other rotational values were determined by speckle-tracking (Echopac, 2D-Strain, GE). For statistical analysis a Student's T-Test was used.

**Results:** In patients with ASD maximal torsion was elevated before intervention in comparison to healthy controls ( $17.8 \pm 8.4^\circ$ ,  $p < 0.05$ ) and dropped significantly after intervention ( $11.3 \pm 5.4^\circ$ ,  $p < 0.01$ ). Similar results were obtained by relief of PST. ( $14.8 \pm 5^\circ$  vs  $10.3 \pm 5.3^\circ$ ,  $p < 0.05$ ). In contrast to healthy children PM-patients had their peak basal rotation before peak apical rotation ( $p < 0.05$ ) resulting in a decreased delay between apical and basal rotation ( $-2.0 \pm 33\%$  vs  $16.9 \pm 14.7\%$  of cardiac cycle,  $p < 0.0001$ ).

**Conclusions:** Curation of RV volume- or pressure-overload by ASD-closure or balloon dilatation of PST is followed by normalization of LV rotational deformation. Electrical abnormal RV cardiac stimulation can influence LV torsion by reversing the timing of basal and apical peak rotation making it an interesting tool for the investigation of dyssynchrony.

#### P-229

##### Benefits for patients with CHD in 3.0 Tesla MR Imaging with MultiTransmit Technology – First Experience

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**Introduction:** Drawbacks of conventional 3.0 T MRI are related to pronounced dielectric shading and SAR limitations and restricts its use in clinical routine work. Almost no information is accessible concerning cardiac examinations. Aim of the investigation was to estimate the impact of MultiTransmit technology (TX) to improve examination strategies in pts with complex CHD and optimization of scanning parameters.

**Results:** As shown in the figure (upper row) enhanced image uniformity was achieved applying two independent RF sources resulting in consistent SAR distribution (right). The reduced SAR allows either the acquisition of more slices per time unit or a reduction of scanning time by about 30–40% in TSE black blood sequences. In SSFP sequences TX decreases the repetition time (actual case: 3.3s to 2.9s) and echo time (1.65s to 1.45s). In addition to an improved image quality (lower right) breathhold acquisition time was reduced.

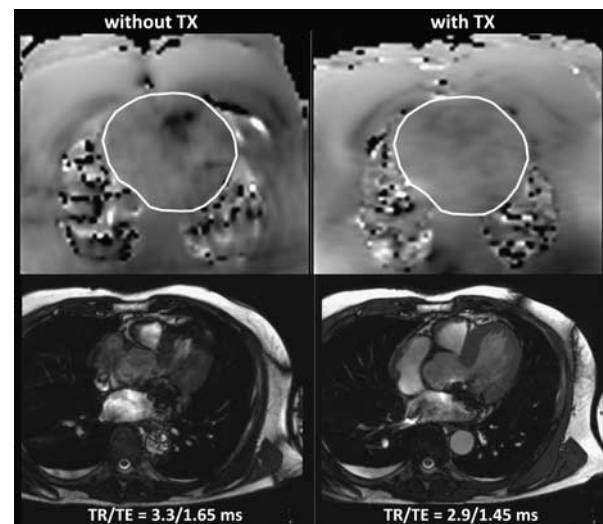


Fig: B1 calibration and example of prosthetic mitral valve using SSFP with and without Tx.

**Conclusion:** MultiTransmit technology speeds up examination of cardiac patients and improves image quality.

#### P-230

##### 3D Wall Motion Tracking: Our Experience in Healthy Children and Congenital Suprarenal Hyperplasia

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**Introduction:** New techniques such as 3D Speckle Tracking (3DST) claim to be the future gold-standard in non-invasive ventricular function diagnosis in children. Our goal is to test their reproducibility and value in a healthy paediatric population and compare it to a probable high-risk cardiovascular population.

**Methods:** 37 patients (age range 4–19 years), 14 healthy children and 23 diagnosed with Congenital Suprarenal Hyperplasia (CSH) on long-term corticosteroid treatment with no structural heart disease were included prospectively. An Artida Toshiba ultrasound with PST-25SX probe was used. After calculating

Ejection Fraction by M-Mode (EFM), 3DST images were acquired for analysis by 2 observers. Image quality score (low, medium and high) was assigned. Comparative inter-observer statistic analysis was done with Lin's correlation coefficient on: EF by 3DST (EF3D), Area Tracking (AT) and 3D Strain (3DS). Reproducibility between EFM and by 3DST was analyzed with Passing-Bablok regression line. Inter-group (healthy and CSH) analysis was performed with the same parameters obtained by one observer.

**Results:** There were no significant differences between healthy and CSH children in gender (64.3% vs 60.9% male), mean age (10.6 vs 11.4 years), weight and BSA.

**Inter-observer comparison:** Global mean quality in all 37 patients was medium, 10 (27%) in the high quality group (mean age 11.8 years). Observer 1 mean EF3D, AT and 3DS were 43.1%, -27.1% and 35.4% respectively. Observer 2 were 45.6%, -27.8% and 42.8%. Global Lin's coefficient agreement was 0.39, 0.29 and 0.33 (low) for EF3D, AT and 3DS respectively. In the high quality group 0.44, 0.42 and 0.61 (acceptable). Passing-Bablok regression line revealed no differences only in the high quality group.

**Intergroup data:** In healthy and CSH children Mean EF by M mode were 67.7% and 65.3% ( $p = 0.25$ ).

EF3D were 49.7% and 43.1% respectively ( $p = 0.002$ ), mean AT were -31.4% and -25.6% ( $p = 0.002$ ) and mean 3DS were 43.6% and 42.2% ( $p = 0.76$ ).

**Conclusions:** These preliminary data in 3DWMT show an acceptable agreement among observers in high quality studies only. Assuming these limitations, we describe that EF3D was lower than M mode standard in all patients, and that EF3D and AT are significantly lower in CSH patients compared to healthy ones.

### P-231

#### Left ventricular myocardial function in ALCAPA: a speckle tracking study.

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**Introduction:** ALCAPA provides a unique opportunity to study coronary induced ischaemia in the paediatric population.

**Aims:** To investigate changes in regional wall motion abnormality in ALCAPA patients pre and post revascularisation using speckle tracking.

**Methods:** Single tertiary centre prospective analysis of pre and most recent post-operative radial, circumferential and longitudinal deformation and (T2P) time to peak deformation (synchronicity of segments), from echocardiograms 2005-10.

**Results:** Eighteen children (8M/10F) underwent surgical re-implantation (1 death). Median age at surgery was 5 months (range 0.03-8.8 yrs). Median follow up was 39 months (range 1-48). ECG ST changes were seen in 75%. Mean fractional shortening was 21% preop with importantly 4 cases being normal (range 6.3-39.6%) and 25.5% post op (range 4-43.7%). Mitral regurgitation and appearance of endocardial fibroelastosis was universal. There was a significant change in global radial ( $p = 0.05$ ) and longitudinal deformation ( $p = 0.05$ ) but not circumferential. There were statistically significant changes between pre and post op echocardiograms in some but not all segments. Significant changes occurred in peak longitudinal strain in the basal septal ( $p = 0.02$ ), mid lateral ( $p = 0.006$ ) and basal lateral segments ( $p = 0.03$ ). There were significant changes in radial peak strain in the lateral ( $p = 0.03$ ), posterior ( $p = 0.02$ )

inferior ( $p = 0.004$ ) and septal segments ( $p = 0.04$ ). No significant changes occurred in any segment in peak circumferential strain. Synchronicity improved post-operatively with ICC increasing for T2P for all 3 fibres (ICC preop vs post op 0.7 vs 0.8 for longitudinal, 0.8 vs 0.9 for circumferential & 0.7 vs 0.9 for radial fibres). Surprisingly there was an increase in T2P for all segments for all 3 fibres types post-operatively. Intra and interobserver variability was low. Patients with normal 2D and M mode pre-op echos had abnormal speckle tracking analysis of the same scans.

**Conclusions:** Contractility (Strain) improved after revascularisation but not in all segments. Dyssynchrony (T2P) improved after revascularisation but not necessarily in corresponding segments. These changes may be due to scar formation. Speckle tracking is a useful technique to assess RWMA, providing better indication of detailed LV systolic function than 2D or M mode.

### P-232

#### Congenital heart defect specific volumetric data in children with Hypoplastic Left Heart Syndrome measured by CMR

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**Background:** For each congenital heart defect (CHD) knowledge of morphological and functional variations is crucial in order to assess an individual case and initiate specific therapy. CMR-based values exist only for left ventricular (LV) and right ventricular (RV) volumes of children and adults with normal anatomy. Therefore, we sought to provide values for RV volumes, mass and function in children with Hypoplastic Left Heart Syndrome (HLHS) in order to be able to consider the defect specific variation.

**Methods:** 57 children ( $5.5 \pm 2.8$  years) with HLHS were evaluated after surgical completion of Fontan-Circulation in a single center. Children with hemodynamic relevant lesions such as tricuspid- or neo-aortic-regurgitation as well as aortic stenosis were excluded. All examinations were performed in freely breathing deeply-sedated children. RV volumes and mass were measured in the end-systolic and end-diastolic phases on fast field echo cine images (TR/TE/ $\alpha = 1,1/1,6/60$ ; FOV:  $240 \times 260$ ) acquired in a short axis planes as a stack of at least 10 contiguous, parallel slices covering the full length of the ventricle.

**Results:** Average volumetric data (mean  $\pm$  SD) are summarized in the table below. The end-systolic (ESV), end-diastolic (EDV) and stroke (SV) volume, as well as the myocardial mass (MM) were indexed to body surface area. No significant gender specific difference was found in young HLHS patients.

RV-EDV ml/m <sup>2</sup>	RV-ESV ml/m <sup>2</sup>	RV-SV ml/m <sup>2</sup>	RV-EF %	CL l/min	RV-MM g/m <sup>2</sup>
73 $\pm$ 23	38 $\pm$ 17	38 $\pm$ 11	52,0 $\pm$ 9,8	3,0 $\pm$ 1,0	73 $\pm$ 26

Compared to normal values from healthy children drawn from the literature and from our own data base, children with HLHS have a higher myocardial mass ( $p < 0,001$ ) and larger end-systolic volume of the systemic ventricle ( $p = 0,0013$ ). All remaining parameters, especially the end-diastolic volume ( $p = 0,04$ ), ejection fraction (EF,  $p < 0,0001$ ) and the cardiac index (CI,



$p < 0.0001$ ), are significantly smaller. Intra- and inter-observer variability was in the range of 4% and 9% respectively.

**Conclusion:** CMR data on ventricular volumes and their variability specific to patients with HLHS have excellent inter-observer variability and fill an important void in the current literature. These reference values allow evaluation of an individual patient to assess his CHD. Additionally, the data can serve as a basis for longitudinal studies, as the time course of systemic ventricular remodeling after surgical completion of the Fontan-Circulation has important implications for patient outcomes.

#### P-233

##### **The pulsed Doppler and Tissue Doppler derived E/E' ratio is significantly related to invasive measurement of ventricular end-diastolic pressure (EDP) in biventricular rather than univentricular physiology in patients with congenital heart disease (CHD)**

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**Objectives:** The value of conventional non-invasive Doppler parameter to predict ventricular end-diastolic pressure (EDP) and diastolic function in congenital heart diseases is limited. The aim of our prospective study is to investigate whether the ratio of mitral early blood inflow velocity to early diastolic velocity of the mitral annulus (E/E') assessed by pulsed tissue Doppler is related to EDP in patients with different congenital heart disease (CHD) undergoing left heart catheterization.

**Methods:** 115 hospital inpatients (64 males) with different CHD referred to cardiac catheterization were simultaneously examined by echocardiography to invasive measurement of ventricular EDP during heart catheterization. The mean age at catheterization was 8.71 years (from 3 days to 18 years). These patients were divided two groups according to the different hemodynamic and morphology: group A included the patients with biventricular heart; group B included the patients with univentricular heart.

**Results:** For all the studied patients, a significant positive correlation was found between E/E' and EDP ( $r = 0.54$ ,  $P < 0.001$ ). EDP correlated rather weak with combined measurements E/global LV early diastolic velocity ( $r = 0.27$ ,  $P = 0.02$ ). A significant relationship was also found between ventricular EDP and early mitral inflow velocity E ( $r = 0.36$ ,  $P = 0.001$ ). The ratio of pulmonary venous flow velocities  $s/d$  was not found to be related to invasive measured EDP ( $r = -0.16$ ,  $P = 0.13$ ). Group A ( $n = 96$ ) had similar result, but for group B ( $n = 19$ ), these parameter did not show relationship to EDP.

The analysis for these parameters showed that the larger area under the curve (AUC) was found for the ratio of E/E' (AUC = 0.77) compared with E/global E (AUC = 0.57). E/E' > 10.7 had 69% sensitivity and 81% specificity for EDP > 10 mmHg.

**Conclusion:** Doppler and tissue Doppler derived E/E' ratio is related to simultaneously invasive measurement of EDP in a heterogeneous group of CHD and may provide an important surrogate non-invasive estimation of ventricular diastolic performance in the routine follow up of these patients, but for some kind of heart defect including univentricular heart, the correlation of E/E' ratio and EDP should be considered carefully.

#### P-234

##### **The novel 2D strain reflects improvement and remodeling of LV function rather the conventional echo parameter after aortic valve repair in pediatric patients**

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**Objectives:** To evaluate the outcome and regional and global left ventricular (LV) function after aortic valve repair in children with congenital aortic valve disease.

**Methods:** 32 consecutive patients aged 1.96 years undergoing aortic valve repair due to valve stenosis (AS group,  $n = 21$ ) or aortic regurgitation (AR group,  $n = 11$ ) were studied over a follow-up time period of 12 month in regard to change and adaptation of myocardial function using conventional and novel echocardiographic methods including two-dimensional (2D) strain echo. Conventional and 2D strain echo studies were performed and analysed off-line using commercially available software (EchoPac 6.1.0, GE).

**Results:** The peak aortic valve gradient decreased from  $62.04 \pm 30.34$  mmHg before surgery to  $22.80 \pm 14.13$  mmHg 2 weeks after surgery and  $35.73 \pm 22.11$  mmHg 12 months after surgery ( $p = 0.01$ ). The degree of AR was decreased significantly to grade 0 in 20 children and grade I in 12. There was a significant reduction of thickness of interventricular septum (IVS) and posterior wall resulting in improvement of LV mass index ( $p = 0.007$ ,  $p = 0.043$  and  $p = 0.001$ , respectively). Significant reduction of myocardial thickness was in found especially in the IVS in the AS group ( $p = 0.008$ ) and the significant reduction of LV end-diastolic dimension (EDD) was found in the AR group ( $p = 0.007$ ). 2D strain analysis demonstrated that the global peak strain, global systolic strain rate and global early diastolic strain rate improved significantly for all the patients during the study period after aortic valve repair ( $p < 0.001$ ,  $P = 0.037$  and  $P = 0.018$ , respectively). The global strain and strain rate correlated significantly to IVS thickness ( $r = 0.002$  and  $r = 0.003$ ), LV mass index ( $r = 0.02$  and  $r = 0.015$ ) and EDD ( $r = 0.26$  and  $r = 0.005$ ).

**Conclusion:** Aortic valve repair surgery in pediatric patients results in improvement of global and regional systolic and diastolic LV parameter, which was demonstrated by the 2D strain parameters rather than the conventional echocardiography.

#### P-235

##### **Assessment of Standard and Non-Standard Echocardiographic Acoustic Windows to Visualize the Right Ventricle in patients with Tetralogy of Fallot**

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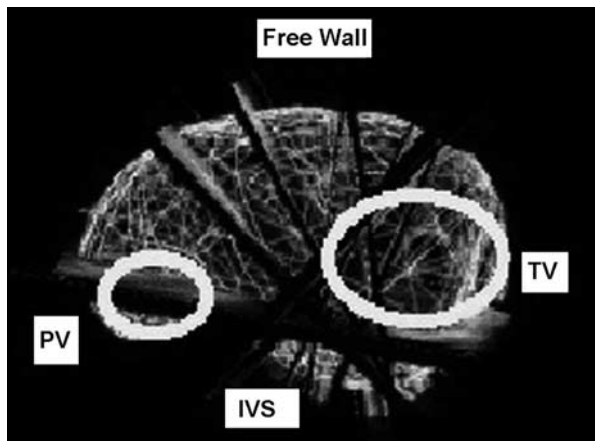
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**Purpose:** The importance of the right ventricle (RV) in assessing patients with congenital heart disease, pulmonary hypertension, heart failure and CAD is increasingly recognized. It is difficult to monitor changes in RV size and function using echocardiography because of difficulty with visualization. The purpose of this study is to evaluate the ASE published guidelines for RV imaging and evaluate alternate non-standard views to visualize the RV.

**Background:** Accurate volumetric information of the RV using 2D or 3D imaging requires complete visualization of the RV. This project evaluates the optimal views for visualization of the



RV and offers additional non-conventional acoustic windows for those with limited acoustic windows.



**Figure 1 - Surface reconstruction of the RV.**  
Each black line intersecting the surface reconstruction represents a 2D image from an apical acoustic window.

**Methods:** Image acquisitions were done in 5 normal and 5 subjects with TOF. Each subject was imaged from multiple acoustic windows including parasternal, apical, right upper sternal border (RUSB), high and low parasternal, subcostal and foreshortened apical. We then superimposed the scan plane from each image on a surface reconstruction of the RV in that patient to assess the position of the scan plane relative to the 3D reconstruction. The reconstructions were done using the Piecewise Smooth Sub-division Surface software developed at the University of Washington and the VentrPoint Medical System.

**Results:** The addition of non-standard acoustic windows improved visualization of the RV. The basal to mid-ventricular inferior lateral wall was the most difficult area to visualize. Low parasternal and foreshortened apical views with rotation around the RV proved to be the most effective acoustic windows to visualize this region as shown in the Figure 1. High parasternal acoustic windows and RUSB provided optimal visualization of the pulmonary infundibulum, PV leaflets and RVOT. Additionally, the 4 chamber view was found to miss the true apex in TOF whereas an oblique view of the apex taken from a low parasternal or oblique foreshortened apical provided optimal visualization of the RV apex in TOF patients.

**Conclusion:** Visualization of the entire RV is critical to assess volume and function. The addition of non-standard acoustic windows allows for more complete visualization of the RV in those patients that are difficult to image, especially those with dilated RV's.

#### P-236

##### **Transcatheter closure of patent ductus arteriosus (PDA): Comparison of Amplatzer duct occluder with the new Amplatzer duct occluder II**

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**Objective:** To assess efficacy and short term results of the new Amplatzer duct occluder (ADO II) compared to Amplatzer duct occluder (ADO).

**Background:** Transcatheter PDA occlusion by ADO device is effective for moderate to large PDA's and new ADO II device may be better in certain types of PDA, and in smaller children.

**Method:** Retrospective case review of occlusion rate, risk of embolisation, and restriction to flow in left pulmonary artery (LPA) or descending aorta after PDA occlusion by ADO or ADO II device. Choice between ADO vs. ADO II was based on duct morphology after angiography.

**Results:** 52 patients (mean age 3 yrs 0.3–16 yrs, mean weight 12.9 kg 4.5–57 kg) underwent antegrade closure of PDA with ADO and 69 patients (mean age 2 years 0.3–11.7 yrs, mean weight 10.3 kg 3.9–45.5 kg) underwent antegrade closure of PDA with ADO II from January 2008 and November 2010. Mean PDA diameter on angiography in ADO group was 2.6 mm and for ADO II group was 2.4 mm. There was immediate occlusion of PDA on angiography in 28/52 (53%) patients in ADO group compare with 43/69(63%) patients in ADO II group. Echocardiography before discharge showed complete closure in 45/52 (87%) patients in ADO group compared to 63/69 (91%) patients in ADO II group. Mean fluoroscopy time for ADO group was 12 minutes (median 7.5 min) and 11 minutes (median 8 min) for ADO II group. Two ADO devices embolised into the descending aorta and two ADO II devices embolised into the pulmonary artery. All were retrieved successfully. Flow restriction into the descending aorta was not seen, but mild LPA flow restriction was noted in 6 of the ADO II group. **Conclusion:** Both Amplatzer duct occlusion devices are safe and effective. The ADO has a greater risk of embolization into the descending aorta than the ADO II. ADO II closure rate is as good as the ADO but is associated with an increased incidence of LPA flow restriction.

#### P-237

##### **Ventricular Septal Defects Closure by Transcatheter Patch in patients with Down Syndrome**

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Surgical or transcatheter closure of perimembranous ventricular septal defects (PMVSD) may result in complete heart block (CHB) in a higher percentage of patients with Down syndrome than in genetically normal subjects. We used Transcatheter Patch (TP, a wireless bioabsorbable device) in 5 patients with Down syndrome and PMVSD. Median defect size was 11 mm. The patches were either small in size (4) or medium (1). They were implanted by either late release method (3) or the immediate one (2). The follow up period was more than 5 years in 3 and around one year in the remaining 2. No patient developed CHB during the follow up. TP may be considered as the first choice in patients with Down syndrome for VSD closure.

#### P-238

##### **Acute Heart Failure after Percutaneous Pulmonary Valve (Melody® Valve) Placement**

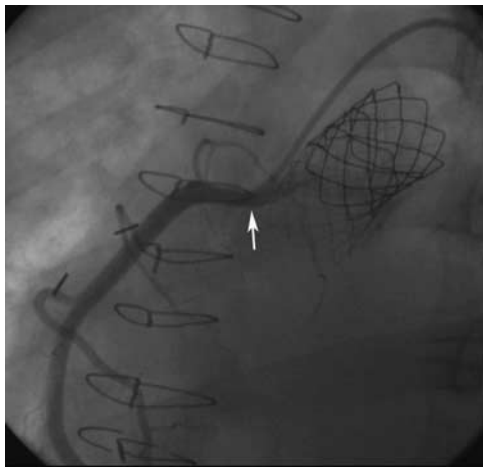
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**Introduction:** We present a case of a 38 year old male with pre-existing left ventricular diastolic dysfunction who developed acute heart failure after Percutaneous Pulmonary valve implantation (PPVI).

**Case Summary:** His operative history consisted of initial subaortic resection at 3 years of age. At 13 years of age, he had placement of a left ventricle to descending aorta graft. At 20 years of age, a Ross procedure, with autograft pulmonary valve to the aortic position and an RV to PA homograft conduit.

Catheterization 10 months prior to admission revealed a 37 mmHg RV conduit gradient. LV end diastolic pressure was 17 mmHg. (Table 1). There was mild pulmonary regurgitation. It was determined that he would be a candidate for PPVI, which was performed 4 months later. Coronary angiography performed with balloon dilation of the conduit showed no risk of coronary compression by valve placement. Implantation of a Melody valve (Medtronic, Minneapolis, MN) was performed without complication. Post-valve placement RV to PA peak systolic gradient was 17 mmHg. Left-sided hemodynamics were not re-measured. Three weeks post-implant, he was re-admitted with progressive symptoms of dyspnea. He had bilateral lung crackles, increased JVD to 8 cm, mild liver enlargement and lower extremity pitting edema. Chest X-ray showed bilateral pleural effusions. Echocardiogram documented good pulmonary valve function with mild stenosis (MIG = 20 mmHg; mean = 12 mmHg) and normal systolic function. Cardiac CT suggested impingement of the Melody stent on the right coronary artery but coronary angiography and cardiac enzyme levels were normal. (Figure 1) LV end diastolic pressure was found to be 45 mmHg (Table 1).



After aggressive diuresis and clinical improvement, he was dismissed home on furosemide 40 mg twice daily and losartan 100 mg.

**Conclusions:** This case illustrates a previously unreported side effect of Melody valve placement in an individual with pre-existing diastolic dysfunction – namely, acute elevation in left ventricular filling pressure due to increased LV preload. filling pressure (4). This case suggests that individuals with RV to PA conduit stenosis and underlying diastolic dysfunction may not experience prompt improvement in symptoms and may in fact have exacerbation of heart failure after PPVI.

Table 1. Invasive hemodynamics before and after PPVI

	06/2009	08/2010		09/2010
		Pre-PPVI	Post-PPVI	
RV systolic	66	69	59	–
RV EDP	13	20	23	–
PA systolic	29	30	42	–
LV systolic	109	100	–	115
LV EDP	17	26	–	45
CI (L/min/m <sup>2</sup> )	2.5	2.4	–	–

Catheter-measured hemodynamics before, during and after PPVI. All pressures are in mmHg. RV = right ventricle; EDP = end diastolic pressure; PA = pulmonary artery; LV = left ventricle; CI = cardiac index.

## P-239

### Interventional catheterisation after total cavopulmonary connection: experience in 68 patients.

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**Introduction:** Total cavopulmonary connection (TCPC) is performed in patients with single ventricle to allow the passive flow of systemic venous blood to the lungs. In this population interventional catheterization is needed to treat residual defects or complications. We discuss our results concerning 68 patients. **Patients and methods:** From January 1995 to December 2010, 68 patients had TCPC. There were 2 early take-down and 5 deaths (7.4%). 3 patients were lost at follow-up, 58 are regularly evaluated. Mean age at TCPC was 5 years (2.5–18); mean interval between TCPC and catheterization 5.6 years (1.5–15). One patient had severe protein-losing enteropathy and one plastic bronchitis. All patients underwent catheterization because of cyanosis or after >10 years from TCPC.

**Results:** 55 catheterizations were performed in 47 patients. 16 patients (34%) evaluated after a median period of 8 years (4–15) from TCPC had low venous pressure, absence of right to left shunt and did not need any intervention. Oxygen saturation was significantly lower in patients requiring interventional catheterization (90.5 + 4.8 and 93.1 + 5.2 and p = 0.003). Interventions, performed in 31 patients (66%) consisted into: fenestration closure (n = 15), embolization of venous vessels prompting right to left shunt (n = 14), stenting or reconnection of pulmonary arteries (n = 4), stenting or reconnection of systemic veins (n = 8), other procedures (n = 3). In two patients the fenestration could not be closed because of high venous pressure. After interventions oxygen saturation increased from 90.5 + 4.8% to 94.73 + 3.6% (p = 0.002). **Conclusions:** Our data show that an unexpected high proportion of patients with TCPC are doing well late after TCPC. However, the majority of patients continue to need interventions generally aimed to suppress stenoses at various levels of TCPC or vessels prompting right to left shunt. This population should enter into a multicenter program aimed to identify patients at risk.

## P-240

### Obstruction of systemic veins: early and medium term results of percutaneous interventions

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**Background:** Few data exist concerning mid term results of interventions for stenoses of systemic veins. We review our experience in patients with and without congenital heart disease (CHD) over a 10 year period.

**Methods:** Since January 2000 to December 2010, 28 catheterizations were performed in 24 patients (12 females) in order to treat stenoses or obstruction of systemic veins. 12 patients had repaired CHD, 2 had had cardiac transplantation, the remaining 10 patients had chronic diseases needing permanent central venous catheters. Catheterization was performed to treat superior vena cava syndrome (n = 16) or to allow access to the heart (n = 12). Mid term results were evaluated by cardiac catheterization or CT scan.

**Results:** Median age and weight of patients at catheterization were 6 years (range 0.1–16) and 15 kg (range 2–60). The affected vessels were: superior caval vein (n = 10), innominate vein (n = 5), subclavian vein (n = 2), inferior caval vein (n = 6), femoral veins (n = 9); 6 patients had multiple lesions. Procedures included 4 simple dilatations and 24 stenting with or without

pre- or post-dilatation. In total 39 stents were implanted. In all but 2 patients, it was possible to treat the lesion. There were 2 procedural complications (8.3%): 1 perforation of superior caval vein leading to cardiac tamponade and one acute stent occlusion without sequelae. Two late deaths occurred, due to CHD. (1) or prematurity (1). At a median follow-up of 3 years (1.5–8) patients were reevaluated with catheterization (n = 12) or CT scan (n = 10). We observed 4 complications of stenting (10%): 2 occlusions, 1 fracture, 3 restenoses.

**Conclusions:** Interventional treatment of obstructed systemic veins offers good immediate results and a low incidence of complications. However, stent thrombosis or intrastent growth raise concern on mid term results.

#### P-241

##### **Dilatation of pulmonary artery stenosis and coarctation of the aorta with new Cobalt-Chromium Stents (Andrastents XL & XXL)**

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**Introduction:** Stenosis of the major vessels are currently treated successfully with stent implantation. Recently new cobalt – chromium stents (namely XL and XXL Andrastents, Andramed, Germany) were introduced into clinical practice. This alloy is combined with high biocompatibility, radial strength and flexibility. The objective of this paper is to present our experience with application of Andrastents XL and XXL in the dilatation of stenosis of pulmonary artery and coarctation of the aorta (CoA).

**Methods:** There were 19 patients treated with 21 Andrastents. In 7 patients (age 16–24 years, weight 54–77 kg) before Melody valve implantation prestenting with 9 Andrastents XL or XXL (length 30,39 or 48 mm) in calcified pulmonary homograft were done. In one patient with long stiff stenosis 3 stents were necessary. In another 9 patients (age 15–55 years, weight 47–85 kg) with native CoA or ReCoA 9 Andrastents (XL or XXL length of 30, 39 or 48 mm) were implanted.

In 3 patients (age 6, 9 and 10 years, weight 17, 30 and 33 kg respectively) 3 Andrastents 30 XL were implanted to the stenosed right or left pulmonary artery (PA) closely to the bifurcation. Mean fluoroscopy time in case of CoA was 7,3 (3,9–13) min and in PA branch stenosis 12,9 (8,7–16,4) min. The follow up ranged from 1,5 to 25 (mean 9) months.

**Results:** All procedures were finished successfully. In follow-up no fracture of the stents were observed. In cases of Andrastent and Melody valve implantation mean gradient in RVOT decreased from 42,8 to 17,8 mm Hg, in case of CoA and reCoA from 48,2 to 11,3 mm and in case of PA branch stenosis from 47 to 19,6 mm Hg. No aneurysm formation was observed in any patient.

**Conclusions:** Implantation of Andrastents XL and XXL are very good therapeutical option for the treatment of stenosed great vessels.

#### P-242

##### **Stent implantation in aortic coarctation in children: ten year experience**

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**Aim:** To report our experience on stent implantation in aortic coarctation (AC) in children.

**Methods and patients:** Between January 2000 and December 2010, 71 consecutive patients aged <18 years with AC had a percutaneous treatment in our institution (29 females; age  $12.5 \pm 3.7$  years; weight  $50 \pm 20$  kgs). Forty six subjects (65%) had a native AC while 25 (35%) had a recurrent AC. (21 post-surgery, 6 post balloon angioplasty). Procedures were performed under general anesthesia. Bare stents (BS) were used in 37 patients, while covered stents (CS) in 34 subjects.

**Results:** Peak-to-peak gradient reduced significantly from  $39 \pm 12$  mmHg to  $6 \pm 3$  mmHg ( $p < 0.001$ ). AC diameter increased significantly from  $6 \pm$  mm to  $14 \pm 4$  mm ( $p < 0.001$ ). Fluoroscopy time was a median of 16 minutes (range 7–48 minutes). The median balloon to aortic coarctation ratio was 3 (range 1,5–6). Early complications occurred in 6 subjects (8%): death due to rupture of aorta in a 17 years old girl with aortic recoarctation associated with bicuspid aortic valve and aneurysm of the aorta (treated by using a Palmaz BS); artero-venous femoral fistula (1 pt); thrombosis of femoral artery treated by drugs (1 pt); stent embolization (2 pts treated with BSs); epileptic attack in one subject.

Early complications occurred more frequently in BS compared to CS. (11% versus 5%;  $p = 0.04$ ). Subjects treated with CSs had tighter stenosis ( $4.8 \pm 2$  versus  $7 \pm 3$  mm;  $p = 0.001$ ) and needed larger sheath (median 12 versus 11 Fr;  $p = 0.01$ ). There were no differences for other variables. Follow-up was a median of 5,5 years (range 1–10 years). Redilatation was needed due to somatic growth in 20 subjects (10 BS and 10 CS). In four subjects treated with BS another stent (CS) was implanted because of restenosis and development of aneurysm of the aortic wall. Ten CS were redilated successfully 1 to 4 years after the initial procedure. Ten subjects still need anti-hypertensive drugs at latest follow-up.

**Conclusions:** Percutaneous treatment by using stents in aortic coarctation is a safe and effective procedure. Introduction of CS allowed the reduction of risks and the treatment of tighter aortic coarctation.

#### P-243

##### **Transcatheter Closure of Perimembranous Ventricular Septal Defects Using the Amplatzer Perimembranous Ventricular Septal Defect Occluder in Children: Initial and 4-year Results. A Multicenter Study**

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**Introduction:** In this report we present experience with 65 patients (pts) with PMVSDs who underwent transcatheter closure at 5 different Institutions with the Amplatzer PMVSD occluder and had completed a 4-year follow-up.

**Methods:** The age of the pts ranged from 0.3 to 15 years and the weight from 5 to 46 Kg. During the study period 24 (38%) other patients were excluded from transcatheter closure because they did not fulfil the patient selection criteria (distance less than 2 mm from the PMVSD to the aortic valve, size of VSD in relation to patients age).

**Results:** The device was permanently implanted in all patients. Complete occlusion of the communication at six month, one-year, and 2-year follow-up was observed in 89.2%, (58/65 pts), 92.3.8% (60/65 pts), and 93.8% (61/65 pts). Main complications included: Early. a. Device embolization (2 patients-catheter removal), b. severe procedural bradycardia (4 pts) and Mobitz II heart block (3pts, sinus rhythm after device removal). Late. Complete heart block (1 pt-epicardial pacemaker was inserted).



No other patient developed heart block during the 4-year follow-up. Three patients developed a mild aortic regurgitation not seen early after the procedure. In 2 of them the regurgitation was not seen at the 1-year follow-up. No other complications were observed.

**Conclusions:** Transcatheter closure using the Amplatzer PMVSD occluder is as a safe and effective nonsurgical alternative that should be offered in properly selected patients with PMVSDs. However, due to anatomic reasons, this therapy cannot be offered to significant number of patients with these defects.

#### P-244

##### **Percutaneous closure of atrial septal defect with Biodegradable atrial septal occluder in children**

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**Background:** The Biostar septal occluder is a new transcatheter atrial septal defect occlusion device. This study analysis the data to determine the most significant factors is successful use of the device.

**Methods:** From October 2009 to January 2011, 34 patients (18 male) were catheterized to close an atrial septal defects with the Biostar in children. The age of the patients ranged from 2.5 to 13 (mean  $6.7 \pm 3.4$ ) years, the body weight ranged from 11–55 (mean  $22.5 \pm 10.9$ ) kg. Transesophageal echocardiography was performed simultaneously in 6 patients

**Results:** The device was successfully implanted in 93.7% (32/34) of patients, but had to be abandoned in 2 patients, because of deficient aortic rim. In one patient, two Biostar devices were used to occlude two separate atrial septal defects. The mean stretched diameter of the ASD was  $13.1 \pm 3$  mm, the mean device diameter was  $29.8 \pm 3.2$  mm. The devices were implanted in 24 patients with one hole secundum ASD, in 4 patients with 2-hole, in 3 patients 3-hole and in 3 patients with fenestrated ASD. The procedure was unsuccessful in one patient and the device had to be removed. There was one severe complication, because of the device embolization, the patient underwent semiurgent surgical intervention for device removal and ASD closure. The occlusion rates were with the Biostar 77.4% (24/31) after 24 hours, 96.8% (30/31) after 6 month.

**Conclusion:** The Biostar septal occluder is best suited for small to moderate atrial septal defects. Biostar is absorbed and replaced with healthy native tissue; future access to the left atrium may be achieved. Percutaneous closure of ASD ostium secundum type defects with the Biostar is safe and effective with high success rate and excellent mid-term outcome.

#### P-245

##### **NT-proBNP correlated with strain and strain rate imaging of the right ventricle before and after transcatheter closure of atrial septal defects**

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**Background:** Atrial septal defect (ASD) accounts for 10% of all congenital heart lesions.

**Our objectives were:** Using strain and strain rate imaging to assess right ventricular (RV) function in patients with RV volume overload due to ASD and correlate the results with the level of N-terminal pro-brain natriuretic peptide (NT-proBNP) before and after device closure.

**Subjects & Methods:** 45 patients (27 females and 18 males) mean age 21.53 years were diagnosed with ASD and admitted for percutaneous closure. For assessment of systolic RV function by echocardiography, we measured tricuspid annular plane systolic excursion (TAPSE), strain, and strain rate Imaging (SRI). Amplatzer ASD Occluder used under general anesthesia. Plasma levels of NT-proBNP concentrations were measured before and three months after closure.

**Results:** Amplatzer ASD closure was achieved in all patients with no complications. The mean ASD diameter was 15.15 mm and the stretched diameter was 18.72 mm. Our study group had average or mild increased pulmonary artery pressure and mean shunt volume was 2.3. The mean left atrial diameter in pre ASD closure group was significantly higher than the control, with non significant difference after closure; mean left ventricular end diastolic dimension showed non significant difference from either control group or post ASD closure group. While mean right ventricular end diastolic dimension showed markedly reduction post ASD closure, and it was significantly higher than control group. Global RV strain and peak systolic strain rate (PSSR) were significantly higher in ASD group than control group. Three months after device closure, there was significant reduction of the global RV strain, PSSR, TAPSE and mean NT-proBNP levels. The NT-proBNP levels found to be correlated with pulmonary artery pressure, TAPSE, as well as the global RV strain and PSSR. **Conclusion:** Amplatzer ASD occluder is simple in construction, easy to deploy, and can be withdrawn and repositioned many times. Volume overload induced by ASD is associated with increased RV strain values, which return to normal after closure. NT-proBNP is a parameter which correlates to RV function, pulmonary artery pressure and the amount of interatrial shunt volume caused by the underlying ASD.

#### P-246

##### **Follow-up results of transcatheter pulmonary valvotomy in patients with pulmonary atresia and intact ventricular septum**

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**Objectives:** We investigate the long term outcomes of patients with pulmonary atresia and intact ventricular septum (PA-IVS) following transcatheter pulmonary valvotomy.

**Methods.** Between 1995 and 2010, 55 neonates with PA-IVS and tricuspid valve Z-score  $> -3.5$  (ranging from  $-3.5$  to  $0.5$ , mean  $-1.1 \pm 0.9$ ) underwent attempted transcatheter decompression of right ventricle. A radiofrequency guidewire or coronary guidewire was used for perforation of pulmonary valve. Balloon dilation was subsequently performed.

**Results:** Of the 55 patients, perforation of atretic pulmonary valve was attempted with a radiofrequency guidewire (PA 120) in 49 and with a guidewire in 6. The procedure was successful in 49 patients: 4 with a guidewire and 45 with a radiofrequency guidewire, but failed in 6. Three of the 6 failure cases were complicated with pericardial effusion. Of the 49 patients, the mean right ventricular systolic pressure decreased from  $115 \pm 22$  to  $54 \pm 12$  mmHg following valvuloplasty. ( $P < 0.01$ ) A stent was implanted to maintain patency of ductus in 4. Nine underwent a right ventricular outflow tract (RVOT) patch with or without a shunt and 2 underwent a shunt because of persisted cyanosis requiring PGE1 and/or development of infundibular stenosis. Ligation of ductus was performed in 4. There were 4 early mortalities: 1 sepsis & 3 heart failure. There was two late



mortalities: severe right heart failure in 1 and renal failure in another. The mean tricuspid valve Z score was significantly lower in patients with RVOT patch than those without. ( $-1.99 \pm 0.34$  vs.  $-0.84 \pm 0.85$ ,  $P < 0.05$ ) In the most recent follow-up, 41 patients achieved biventricular circulation and 2 had 1.5-ventricle circulation.

**Conclusions:** Transcatheter pulmonary valvotomy can be a definitive treatment in selected patients with PA-IVS. Those with a significantly hypoplastic right ventricle may require RVOT patch despite a successful pulmonary valvotomy.

#### P-247

##### **Re-Interventions in pulmonary arteries during the classical Fontan palliation pathway**

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**Objective:** The pulmonary vascular bed is of great importance for the univentricular circulation on the long run. Pulmonary arteries in Fontan patients may be affected by previous shunts and bandings, they may kink or being compressed through aortic arch reconstruction. This study wants to review the prevalence of both surgical and catheter based treatment of pulmonary artery obstructions in all Fontan patients treated in our institution between the first operation and six months after TCPC.

**Methods:** From 10/2000 to 12/2010 123 consecutive patients followed the classical Fontan pathway in our institution. The diagnoses were HLHS in 57, other left-sided obstructions requiring Norwood or Damus-Kaye-Stansel in 22 and HRHS in 34 patients. PAB was done in 11 patients, but not in advance of hybrid procedures.

A RMBTS was placed in 45, a Sano-Konduit in 38 patients. Bidirectional Glenn was performed between 2,5 and 5 months, extracardiac Fontan with a median body-weight of 14 kg (10–17). All but 13 patients received a 4 mm fenestration, which was closed interventionaly six months later, whereas the unfenestrated group underwent diagnostic angiography. Surgical charts and catheterization data were retrospectively analyzed.

**Results:** 17 patients (13,8%) had to be treated for pulmonary artery stenosis (LPA: 15, RPA: 2). 5 patients underwent surgical PA patch-enlargement during Glenn or TCPC, all of them received stents later on. 12 patients were treated interventionaly, the RPA obstructions (previous BTS) by balloon-dilatation, the LPA cases (compression by reconstructed aortic arch) with stenting. Median age at treatment was 54 months (2–105), median age at last catheterization 7,8 years (3,2–31). Only 6 procedures out of routine were necessary.

**Discussion:** Obstructions of the pulmonary vessels may impair the in itself reduced growth potential of the vascular bed and may lead to sequelae like PLE and plastic bronchitis. Repeated interventions are necessary in selected cases to provide good vessel size till adulthood. The fact, that the vast majority of our group (86%) did not show any relevant obstructions may be promising and set in comparison to new hybrid strategies in the future.

#### P-248

##### **Right heart failure due to an acute obstructing fracture of a Melody<sup>®</sup> valve stent**

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**Introduction:** Transcatheter pulmonary-valve-implantation has become a well established treatment of conduit dysfunction. Wire fractures, especially when no pre-stenting was done may occur, usually not causing functional problems. We report a case of an acute obstruction due to stent fracture two years after PPVI.

**Patient:** A 21 years old male patient with Fallot who underwent VSD closure and RV/PA conduit with 12 years received a 20 mm PPV in May 2007 without pre-stenting. Chest X-ray revealed a first singular wire fracture after three months. During FU over two years more fractures occurred but with no influence to the stent integrity. Cardiac MRI 6,12 and 24 months after implantation showed normal stent morphology and valve function.

Patients case was discussed on the Implanter Meeting 2009 and close observation was recommended.

29 months after implantation the patient was admitted with acute right heart failure. Because of a “funny” shape of the PPV in chest X-ray he was referred to the cardiology department and underwent acute cardiac catheterisation.

The anterior proximal circumference of the PPV had completely lost its integrity with the fractured part protruding into the lumen of the distal portion in every systole causing subtotal obstruction.

The lesion could be passed to reestablish adequate pulmonary blood flow. Three prestents (Intrastent LDMax<sup>®</sup> 20 × 36 mm) where implanted excluding the broken PPV. Finally a second Melody<sup>®</sup> on a 20 mm Ensemble<sup>®</sup> could be placed successfully. No residual gradient and full competence of the valve was observed during the follow up period of 14 months. No wire fracture could be found so far, although detection is difficult in an ensemble of five stents.

**Discussion:** Being a covered stent the Melody<sup>®</sup> valve may obstruct itself when broken parts – fixed by the tissue – are pressed into the lumen by the force of blood flow. The case history stresses the recommendation for pre-stenting every conduit before implanting a Melody<sup>®</sup>. Early treatment with a new PPV in not pre-stented patients may avoid severe complications.

#### P-249

##### **Use of a Telescopic System for Transcatheter Radiofrequency Perforation and Balloon Valvotomy in Infants with Pulmonary Atresia and Intact Ventricular Septum**

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**Objectives:** to evaluate the impact of a new technique for transcatheter radiofrequency (RF) perforation and valvotomy on required fluoroscopy time, early morbidity and mortality in newborns with pulmonary atresia and intact ventricular septum (PA-IVS). PA-IVS is a complex congenital heart disease with great morphological variability. Approximately two thirds of affected infants may be suitable for transcatheter pulmonary valvotomy. However this procedure remains technically demanding even in experience hands. We reviewed our experience with the telescopic system and we compared early results of two different percutaneous approaches. **Methods:** during the last 10 years 28 infants with PA-IVS underwent RF perforation of the pulmonary valve (PV) and subsequent balloon dilation at our Institute. In all cases cardiac catheterization was carried out under general anaesthesia, the femoral vein was percutaneously cannulated using a 5Fr sheath and right and left ventricle angiographies were performed. In the first 14 infants we used a 5 Fr Judkins right coronary catheter which was manoeuvred

directly underneath the atretic PV (Group A). Lately a telescopic system consisting of Northstar Lumax Flex and White Lumax Guiding Catheters (Cook) was adopted to obtain the proper position under the atretic PV (Group B). After successful RF perforation of the PV a 0.014 inch super floppy guide wire was advanced into the descending aorta and progressive balloon dilations were performed. Results: there were no significant differences in patient characteristics (age and weight) and pre-interventional echocardiographic findings (tricuspid Z value, right ventricle morphology) of the two groups. The procedure was successful in all but one patient. Procedural morbidity and required fluoroscopy time were significantly lower in Group B. (3 vs 0 and  $44 \pm 17$  min vs  $24 \pm 18$  min, respectively) ( $p < 0.01$ ). Two premature infants of Group A died early; 8 patients of Group A and two of Group B required a mBT shunt.

**Conclusions:** neonatal percutaneous RF perforation of the PV could be effective with a low risk of mortality and morbidity in almost all infants with PA-IVS and patent infundibulum. The use of a telescopic catheter can significantly decrease the incidence of procedure related complications and the fluoroscopy time needed for this intervention.

#### P-250

##### **Morphology of patent ductus arteriosus; a predictor of the outcome of stenting in duct-dependent pulmonary circulation an experience in children hospital, cairo University, Egypt**

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**Background:** Ductal stenting is an alternative to conventional shunt surgery in duct-dependent pulmonary circulation as it avoids thoracotomy. It is preferable now in Egypt as there is waiting list for this critical surgery and sometimes the pulmonary arteries are too small for Blalock-Taussig shunt. The key points for successful PDA stenting depend on the morphology of the duct.

**Objectives:** To evaluate the impact of ductal morphology and heart anatomy on PDA stenting in neonates & young infants with duct-dependent pulmonary circulation in Children Hospital, Cairo University. To correlate the morphology of PDA with the outcome of the stenting procedure.

**Patients & Methods:** During the period from April 2008 until November 2010, 46 patients with duct-dependent pulmonary circulation were referred for PDA stenting. In most of the patients, the diagnosis had been established by echocardiography. The role of angiography is to have a detailed evaluation of the ductus arteriosus morphology and size. All patients with duct-dependent pulmonary circulation, patients unable to undergo a shunt operation.

**Results:** Successful PDA stent was achieved in 29 cases (63%) and failed in 16 cases (41.3%) out of 46 patients. Statistical significant rise in the oxygen saturation was observed in the successful group ( $67 \pm 6\%$  versus  $87 \pm 7\%$ ,  $P < 0.001$ ). Mean age of the patients at PDA stenting was  $20 \pm 12$  days (range 1–210 days) and mean weight was  $3.4 \pm 0.3$  kg (range 2.8–6.5 kg). Ten patients had biventricular physiology and 36 patients had univentricular physiology. In horizontal ducts (19 patients) the success was 100%, in vertical ducts (10 patients) success was 40%, while in tortuous ducts (17 patients) success was 35%. PDA stenting was feasible in 50% of the cases with pulmonary branch stenosis.

Pulmonary atresia with intact interventricular septum followed by tricuspid atresia with pulmonary atresia was the best cardiac anomaly for successful ductal stenting.

**Conclusion:** Select properly the cases for ductal stenting and although tortuous ductus and branch pulmonary stenosis are difficult ones, they still might be achievable. Early duct stenting significantly shortens hospitalization and reduces treatment costs of prostaglandin infusion.

#### P-251

##### **17 years follow up after interventional treatment of aortic valve stenosis in newborns and infants less than six month.**

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**Objectives:** To evaluate the long term outcome of balloon valvuloplasty (BVP) in aortic valve stenosis in newborns and infants.

**Method:** Retrospective analysis of 39 patients under six month of age at time of first BVP with a special focus on a subgroup of 26 newborns with critical aortic valve stenosis.

**Results:** The short term results showed a significant reduction of the peak pressure gradient from mean 55,2 mmHg to 31,4 mmHg ( $p < 0,0004$ ) measured in the cathlab after BVP and from 69,9 mmHg to 40,2 mmHg ( $p < 0,00002$ ) measured in echocardiography after intervention. In the newborn subgroup the reduction of the invasive pressure gradient ( $p < 0,0002$ ) and echocardiographically measured gradient ( $p < 0,0001$ ) were similar. Aortic regurgitation (AR) was observed in 3 patients prior to BVP. After BVP AR was classified as mild in 11 patients and as moderate in 7 patients respectively. Mitral valve stenosis (MR) was noted only in neonates ( $n = 12$ ) prior to BVP with a reduction of severity or complete disappearance after intervention. We did not find any significant relationship whether the BVP was performed using retrograde arterial access or antegrade venous access. There was no significant relationship between the diameter of the balloon and the aortic valve either ( $p > 0,2$ ). Nevertheless we noted a higher risk for AR with a ratio of balloon diameter to aortic valve diameter greater than 0,7.

The mid term results showed freedom from reintervention of 82% after one year and 77% after ten years respectively. 88% of the patients in the newborn subgroup needed a second BVP within 6 months after the first BVP. The long term followup showed a freedom from further procedures of 56% ( $n = 22$ ) after 17 years in all patients and 50% ( $n = 13$ ) in the subgroup respectively.

**Conclusion:** BVP in aortic valve stenosis is a save procedure in patients less than six month of age. The pressure gradient can be reduced significantly a can offer viability without signs for congestive heart failure and without need for reintervention. Even in newborns with critical aortic valve stenosis the results are encouraging.

#### P-252

##### **Incidence of atrial fibrillation after percutaneous closure of patent foramen ovale in patients presenting with cryptogenic stroke**

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**Background:** percutaneous closure of patent foramen ovale (PFO) has been emerging as an effective treatment of PFO, although several studies have reported atrial fibrillation (AF) as a not rare

complication of this procedure. However the precise incidence of AF still remains to be determined in these patient population.

**Objective:** the aim of this work is to carefully assess the incidence of AF occurring after transcatheter PFO closure in the population of a single-center, observational, retrospective study. **Materials and Methods:** this study included 31 consecutive patients with device closure of a PFO after a documented stroke or transient ischemic attack. All patients underwent a telemonitoring event loop recording 1 month before and 1, 3 and 6 months after PFO closure. AF after the percutaneous closure was detected by 12-lead electrocardiogram (ECG) and by the analysis of all ECG-tracings sent by patients using the telemonitoring system.

**Results:** over a mean follow-up period of  $8,7 \pm 6,04$  months and a telemonitoring ECG-recording available to patients for an average of  $83 \pm 35,47$  days, AF after percutaneous closure was documented in only 1 (3,2%) patient. This patient had a large PFO. (5–6 mm) associated with a large ASA, and a severe R-to-L shunt before the procedure, that disappeared after the percutaneous PFO closure. Anyway, the same patient presented a history of previous palpitations and an episode of supraventricular tachyarrhythmia (atrioventricular nodal reentrant tachyarrhythmia), effectively treated with slow-pathway ablation 6 years before PFO closure. All the remaining 30 patients presented a normal sinus rhythm, without episodes of atrial arrhythmias.

**Conclusion:** AF has emerged as a complication of percutaneous device closure of PFO. Furthermore, its incidence in patients with cryptogenic stroke and PFO undergoing closure of the interatrial communication compared with patients suffering from stroke without a closure procedure seems to be similar. It also seems to occur more frequently in elderly patients with slightly enlarged left atria. We retain that long-term ECG monitoring may be useful to prevent recurrent events such as thrombus formation or cerebrovascular accidents.

#### P-253

##### Results of interventional procedures in children with aortic arch hypoplasia or stenosis after coarctation surgery

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In aortic coarctation (CoAo) with borderline aortic arch (AoA), surgical treatment is usually related to coarctation itself. In some cases AoA develops inadequately with age or there are postoperative stenotic lesions requiring reoperation or intervention procedures because of systemic hypertension.

The aim of the study was the analysis of the results of interventional treatment of AoA hypoplasia or stenosis in pts after coarctation surgery.

The material consisted of 34 pts after different types of CoAo surgery (age  $x = 1.8$  years), qualified to reintervention between 2000–2010, on the ground of systemic hypertension and results of ECHO studies (postoperative stenosis or hypoplasia of AoA). **Methods:** All the pts underwent angiocardigraphic/hemodynamic studies at the age:  $x = 7.3$  years and – with the exception of 2 (5.9%) – were subjected to balloon angioplasty (BAA) or stent implantation.

**Results:** In 2/34 pts, the procedure was postponed because of gothic shape of the AoA and the need for the stent to cover the origin of all AoA arteries.

**BAA:** Among 16 pts with AoA stenosis (age:  $x = 2.4$  years, body weight:  $x = 12.0$  kg), in 13 (81,2%), the BAA results were satisfactory (Tab. 1), while in 3 the outcome was poor because

of AoA hypoplasia and inadequate body weight of the pts for stenting. Tab.1. Outcome of BAA in pts with satisfactory results

Results of BAA in 13 pts with AoA stenosis	Pressure in ascending Ao (mmHg)	Ø of stenosis of AoA (mm)	Pressure gradient (mmHg)
Before	111.3	3.7	34.2
After	96.6	5.2	15.6
	P = 0.035	P = 0.14	P = 0.01

**Stenting:** In 16 pts (age:  $x = 12.0$  years, body weight: 42.3 kg), the results of stent implantation were satisfactory. Tab 2. Outcome of stent implantation in pts with satisfactory results.

Result of stent implant. in 16 pts with AoA hypopl.	Pressure in ascending Ao (mmHg)	Ø of the narrowest AoA segment (mm)	Pressure gradient (mmHg)
Before	110.5	8.2	20.6
After	101.5	13.0	4.4
	P = 0.051	P < 0.01	P < 0.01

#### Conclusions:

1. Results of interventional procedures in the majority of children with postoperative AoA stenosis are satisfactory.
2. Limitations of stent implantation in AoA hypoplasia except of AoA anatomy are age and inadequate body weight of pts.

#### P-254

##### Balloon valvuloplasty for critical aortic stenosis in neonates – long term follow-up

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**Introduction:** Catheter balloon valvuloplasty (BVP) is a first choice treatment in newborn with critical or severe valvular aortic stenosis (VAS). Critical aortic stenosis was defined if duct dependent systemic circulation was present, or signs of heart failure due to VAS were observed. The aim of the study was to evaluate the short and long term results.

**Methods:** Forty-three neonates (median age: 9 days, range 1–30 days, mean weight: 3.09 kg, male/female:34/9) underwent BVP from 1955 to 2010. Re-intervention (re-BVP or surgery) rate, time to re-intervention, incidence and predictors for early (<4 weeks) and late (>4 weeks) mortality were evaluated.

**Results:** No procedural death occurred. Left ventricular endocardial fibroelastosis (2), hypoplastic aortic or/and mitral valve hypoplasia (2) and left ventricular diastolic heart failure (1) were associated with cases of early mortality (5/43, 11.6%). Among the 38 survivors Norwood procedure was performed in 1 patient. Re-BVP was indicated in 13 cases (34%). Median time to re-BVP was 11.3 months (14 days to 7 yrs). There was no significant difference in aortic anulus/balloon diameter ratio (0.86 vs 1.02) among the patients with and without re-BVP. Surgical valvulotomy was performed in 2 cases due to severe residual stenosis. Ross procedure was indicated in 7 cases because of progressive aortic valve regurgitation. Median time to Ross operation was 4.2 yrs (7 days to 7 yrs). 1 patient died after surgery. Overall 37 patients are alive (95%). Twenty-one patients (57%) remained free from re-intervention during the follow-up.

**Conclusions:** 1. Left sided cardiac comorbidities, especially hypoplastic mitral or/and aortic valve, endocardial fibroelastosis are risk factor for early mortality. 2. Late mortality rate is low after BVP. 3. Re-intervention rate is considerable, due to residual stenosis (early period) and severe regurgitation (late follow-up).

4. Aortic anulus/balloon diameter ratio was not predictive for re-BVP. 5. Males dominate among patients with critical/severe VAS.

#### P-255

##### **Combined X-ray and MRI-guided cardiac catheterisations (XMR): clinical impact in patients with complex anatomy or raised pulmonary vascular resistance**

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**Introduction:** The benefit of combined X-ray and MRI-guided cardiac catheterisations (XMR) in reducing patient radiation exposure has already been established. XMR catheterisations were primarily performed in our institution for assessment of pulmonary vascular resistance (PVR). In the past few years the indications of XMR catheterisations have widened to include assessment of anatomy and function of univentricular hearts, cardiac output measurement during pharmacological stress in liver patients prior to transplantation and MR-guided interventions. We report the results and clinical impact of XMR catheterisations performed in our institution.

**Methods:** Prospective and retrospective review of all XMR catheterisations performed between Feb 2002 and Sept 2010. The procedures were either performed in our hybrid XMR lab or in separate MRI and cardiac catheterisation suites. Ethics and UK regulatory authority approval were obtained prior to commencement of the XMR and MR-guided interventional programme.

**Results:** 174 studies were performed in 156 patients with median age and weight of 4.4 years (range 4 days to 67 years) and 15 kg (range 3–122 kg), respectively. The study group consisted mostly of patients with congenital heart disease (biventricular circulation [n = 108], univentricular circulation [n = 30]). Fourteen patients had liver problems and underwent 19 cardiac output stress studies prior to liver transplantation and 4 patients had normal cardiac anatomy but were catheterised because of suspicion of raised PVR of respiratory aetiology.

The median time from cardiac catheterisation to intervention (medical, catheter or surgical) was 50 days (range 0–208 days). Fifty-seven patients were found to have elevated PVR > 3 WU.m<sup>2</sup> with median of 12 WU.m<sup>2</sup> (range: 3–66 WU.m<sup>2</sup>). PVR was between 3–5.3 WU.m<sup>2</sup> in 31 patients, 5.3–7 WU.m<sup>2</sup> in 7 patients and > 7 WU.m<sup>2</sup> in 19 patients. Of the first group, accurate PVR assessment led to risk stratification and fenestrated, rather than complete closure, of their ventricular or atrial septal defects in eight. Of the 14 liver patients that underwent cardiac output assessment with stress, seven have been transplanted successfully.

**Conclusions:** XMR catheterisation facilitates risk stratification and careful management planning for patients with suspicion of raised PVR, complex anatomy or prior to liver transplantation.

#### P-256

##### **Use of a modern cardiac catheterisation laboratory greatly reduces radiation dose during structural interventions**

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**Objectives:** To compare the radiation dose incurred during common cardiac structural interventions between an old cardiac

catheterisation laboratory and its replacement in the same institution. To provide guide values for radiation emitted during common structural interventions.

**Methods:** A retrospective review of common structural interventions in patients aged up to 18 years at a large congenital cardiac centre between 2000 and 2009. The radiation emitted for each common structural intervention as a group was compared between an old catheterisation laboratory (era one) and its state-of-the-art replacement (era two). Radiation dose was controlled for operator seniority, fluoroscopy time, patient age and study year. Natural log transformations were applied to radiation dose and fluoroscopy time.

**Results:** The total number of structural interventions was 760. The latter era was associated with a dramatic decrease in unadjusted radiation dose for all interventions, this finding persisting after multivariate adjustment. All procedures showed a reduction in radiation dose of at least 71% (p < 0.001). There was no decrease in fluoroscopy time between the eras.

**Conclusions:** Use of up-to-date hardware in the catheterisation laboratory plays a vital role in minimising radiation dose. We provide guide values for the radiation dose for common interventions.

#### P-257

##### **Innovation: Catheter Interventions in Congenital Heart Disease without Catheterization Laboratory Equipment, The Chain of Hope Experience in Rwanda**

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**Introduction:** Thousands of children live in developing countries with untreated but correctable congenital heart disease and most of them will die due to the lack of suitable medical and surgical facilities. We report the feasibility and safety of cardiac catheterization in a developing country without access to a regular cardiac catheterization laboratory. The equipment used for imaging consisted of a mono-plan C-Arm X-ray system (Siremobil<sup>®</sup> compact, Siemens) usually used by orthopedists, and a portable ultrasound machine

**Method:** During a week, for 4 consecutive years (2007–2010), a team of 2 paediatric cardiologists, 1 anesthetist, 2 nurses and 1 technician from the Chain of Hope-Belgium ([www.chaine-espoir.be](http://www.chaine-espoir.be)) performed cardiac catheterisation at King Faisal Hospital-Kigali, to treat 47 patients (45 children – 2 adults). This team was working with local personnel (doctors, nurses and technicians) to facilitate the transfer of expertise. At the same moment and in the same hospital, an Australian surgical team was performing open heart surgery (<http://rwanda.ooh.org.au/>), providing a surgical back-up in case of catheterization complication.

**Results:** Treatment, using usual guide-wires and catheters for cardiovascular access, was successful in 45 out of the 47 patients, consisting in 29 patent ductus arteriosus occlusions with coils (n = 6) or Amplatzer device (n = 23); 3 secundum ASD closure, 12 pulmonary valve stenosis (PS) dilatation (3 critical PS, 1 PS in a Fallot patient, 2 PS in patient with ASD device closure), 3 dilatation of aortic coarctation. Two cases required surgery, 1 ASD closure after embolisation of an ASD device, 1 PDA to large for device closure. No other complication was observed and most of the patients were discharged from hospital the day after cardiac catheterization with good evolution on follow-up

**Conclusion:** Cardiac catheterization can be performed safely and is highly effective in a country with limited resources. This mode



of treatment is possible without the support of a sophisticated catheterization laboratory. Working with local staff allows the transfer of expertise.

#### P-258

##### **Midterm follow up of interventional closure of atrial septal defect using the Solysafe™ Septal Occluder – Diagnostic impact of fluoroscopy for complication management**

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**Objective:** In August 2010 sales and distribution of the Solysafe Septal Occluder has been immediately stopped for interventional closure of secundum type atrial septal defects (ASD II) because of frequent reports of wire fractures. Therefore, we analysed the incidence of irregularities associated with implanted occluders during midterm follow up.

**Methods:** We evaluated the midterm follow up of all patients after implantation of Solysafe Septal Occluder focussing on the results of fluoroscopic assessments as recommended by the company.

**Results:** Between April 2007 and June 2010 in 51 pediatric patients (male:female = 24:27) at an age (mean  $\pm$  SD) of  $8.0 \pm 4.6$  years (range 1.6–17.8 years) and a body weight of  $29.1 \pm 18.1$  kg (9.5–86) Solysafe Septal Occluder were successfully implanted. ASD II presented as solitary defects (n = 39), two defects (n = 11), or multiple defects (n = 1) with a significant left to right shunt Qp to Qs  $1.7 \pm 0.7$  (1.0–4.0), native resp. balloon stretched size of  $10.0 \pm 3.9$  mm (5–20) resp.  $12.5 \pm 4.7$  mm (6.5–23) determined by transesophageal echocardiography. During midterm follow-up of  $2.1 \pm 0.9$  years (0.5–3.7) patients were clinically asymptomatic and echocardiography revealed a complete closure rate of 94.9% (48 of 51 patients). Fluoroscopy showed in two patients (3.9%) new complications, not determined by echo before: In one patient (age 7.9 years, 25 mm device, no residual shunt) fixed fracture of one wire loop not leaving the plane of the atrial septum, in the second patient (age 5.0 years, 30 mm device, residual shunt) multiple fractures of wire loops on both sides of the device, with embolization of wire fragments to left and right pulmonary artery. Due to fractured wire ends sticking out of the septum and by this injuring the anterior mitral leaflet, cardiac surgery was performed with complete explantation of the device and successful mitral valve repair and uneventful postoperative course. **Conclusions:** Despite low periprocedural complication rate, during midterm follow-up the rate of fractured wire loops of Solysafe Septal Occluder should lead to regular fluoroscopic controls at least every year as recommended by the company. Large size of the occluder and residual shunts were risk factors for complications in our cohort.

#### P-259

##### **Cyanosis due to right-to-left interatrial shunt without pulmonary artery hypertension. Transcatheter occlusion in 63 consecutive patients**

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Interatrial right-to-left shunt responsible of hypoxemia may have a significant impact on exercise tolerance. We report here one centre experience in transcatheter correction of such disease.

From August 1995 to 2009, 63 consecutive patients (37 females and 26 males, mean age  $54 \pm 24$  years), underwent transcatheter closure of interatrial RL shunt because of cyanosis and shortness of breath at exercise. Associated malformations were complex cyanotic heart disease (n = 4), Ebstein anomaly (n = 1), RV hypoplasia (n = 1), pectus excavatum (n = 2), aortic aneurysm (n = 1). Ten pts had a past history of stroke. A real platypnea-orthodeoxia syndrome was noticed in only 17 pts. Majority of patients had cardiac catheterization under local anaesthesia with a sole fluoroscopic control. None of them had pulmonary artery hypertension. Transcatheter closure was performed with a Sideris device (n = 10), a PFO Amplatzer occluder (n = 40), an ASD Amplatzer occluder (n = 8), a Cardioseal device (n = 3), a VSD Amplatzer occluder (n = 2). Device implantation succeeded in all but two (1 Sideris and 1 Cardioseal device). The fluoroscopic time was  $12 \pm 8$  minutes.

All patients had better clinical tolerance after closure with an oxygen saturation  $>92\%$ . All underwent serial echocardiographic follow-up including contrast study. One month after implantation, no shunt was noticed in all but 6 pts (tiny RL shunt). In 5 of these remaining pts, the 6-month control did not show any residual shunting.

Transcatheter closure of interatrial RL shunt responsible of cyanosis is an effective and a safe method. Many devices using a double disk can be employed and provide usually excellent results. The classic platypnea-orthodeoxia syndrome is infrequent and observed in about 25% of this population.

#### P-260

##### **Midterm follow-up after interventional closure of atrial septal defects using the Solysafe septal occluder**

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**Objective:** The Solysafe septal occluders (Swissimplant®) is a self-centering device for interventional closure of secundum type atrial septal defects and patent foramen ovale which was first successfully implanted in 2005. Individual reports of wire fractures terminated the distribution and implantation of Solysafe septal occluders (SSO) in August 2010. Therefore, we contacted and evaluated all patients who underwent interventional closure of an atrial septal defect using this device at our institution.

**Methods:** From 7/2008 to 7/2010 interventional closure of an atrial septal defect using a SSO was performed in 23 patients (18 females, 5 males). Mean age at the procedure was 8.0 years (2.9 to 16.2 years), the native defect size was mean 11 mm (5 to 16 mm) measured by transesophageal echocardiography. SSO types 15, 20, and 25 were used in 7, 13, and 3 patients, respectively. No SSO types 30 or 35 were used. Between 09/2010 and 12/2010 all patients were re-evaluated carefully. In addition to transthoracic echocardiography and electrocardiogram the integrity of the wire frame was controlled by fluoroscopy.

**Results:** Mean 389 days (69 to 799 days) after the interventional procedure all patients were asymptomatic. Echocardiography revealed a flat device in all patients without residual shunt. No wire fractures were detected by fluoroscopy.

**Conclusion:** Thorough check-up revealed no wire fracture or other device related problems in our patients treated with SSO types 15 to 25. However, mean follow-up was only 1 year. Maybe larger devices or devices implanted in persistent foramen ovale are more susceptible to the reported wire fractures. Further careful follow-up evaluation is necessary.

**P-261****Right ventricular outflow tract stenting in very low birth weight newborns**

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**Background:** Correction of neonatal pulmonary atresia with ventricle septum defect (PA/VSD) depends on development of pulmonary arteries. Very low birth weight newborns (VLBW) demand individual strategies to achieve appropriate growth of pulmonary arteries.

**Objective:** We report about our experience in VLBW with PA/VSD between 1,2 and 1,4 kg weight.

**Methods:** Clinical, echocardiographic and angiographic data were reviewed for four patients who underwent six right ventricular outflow tract (RVOT) stenting procedures from June 2008 to February 2010.



**Results:** All patients were interdisciplinary discussed before deciding for intervention. Antegrad pulmonary artery perfusion was preferred to avoid systemic run-off as seen by stenting of the duct. 4 patients underwent stentimplantation in the RVOT. All stents used were coronary artery cobalt-chromium stents (diameter 4 to 5,5 mm). Three patients had no major complication during or after undergoing intervention. Two patients had atrial flutter due to mechanical manipulation in the right atrium which could have been terminated without electrical cardioversion. One patient died 12 hours after intervention due to right coronary artery compression. Time interval between intervention and surgery ranged from 2 to 4 months. The three survivors had been surgically corrected between 3,2 en 5,1 kg weight. Median diameter of pulmonary artery trunk increased from 3,8 mm to 8,2 mm at moment of surgery. Resection of the RVOT stent was possible in all cases and demanded extended resection of infundibular myocardium.

**Conclusions:** Our experience in RVOT stenting is based on a non-representative and small number of VLBW newborns. RVOT stentimplantation in patients below 1,5 kg is technically feasible. Balance between ethical aspects and feasibility in such high-risk procedures in critically ill newborns is essential.

**P-262****Pulmonary Atresia and Intact IV Septum (PA intact IVS) after Successful transcatheter Valvotomy. Impact in right ventricular growth. Additional devices and surgical procedures in 10 years follow up**

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**Introduction:** PA intact IVS after successful Transcatheter perforation Valvotomy (TPV) using radiofrequency/guide wire perforation followed by balloon dilatation of the imperforate pulmonary valve has created a group of patients depending on a number of additional Eco/Angio procedures to assure the good development of the RV besides the implantation of intracardiac devices to treat residual shunts or pulmonary branch stenosis and second Pulmonary valvuloplasty besides surgical procedures.

**Methods and Results:** In the last ten years we studied and follow up 25 selected patients with PA intact IVS. after successful TPV. 2D EcoColorDoppler were used as the control method in outpatient. A total of 55 Cardiac Catheterizations were performed. Intracardiac devices were implanted in 14 + 2 patients using a single Stent in the Ductus. In 3p R-L atrial shunt disappeared after ASD closer with an Amplatzer device. Two Pulmonary artery stents were implanted for pulmonary branch stenosis.

**Surgical procedures:** 6 B-T shunts and 2 Stents removal. One Glen anastomosis. 1 Pulmonary valvotomy and 1 RV Infundibulotomy. Two Surgical ASD closure and 1 B-T shunt closure. 1 Ductus closure.

Right ventricular growth along the somatic growth was seen by Eco 2D and RV angiography although apical absence was still present. The original tricuspid regurgitation disappeared in all. RV function was good. However mild to moderate pulmonary regurgitation was present in 23 p remaining patients.

**Conclusion:** Right ventricular growth occurs after TPV. Tricuspid regurgitation usually disappeared. Stent occlusion may take place after 6–8 months after implantation however in most patients remain open with small degree of shunt, not requiring closure.

Pulmonary branch Stenosis related to a B-T surgical shunt can be release by implanting a branch stent.

Although a number of additional procedures are required in this group the clinical situation is satisfactory in functional class I. The only doubt will be the future evolution of the pulmonary regurgitation.

**P-263****Interventional treatment of aortic coarctation in neonates and infants – one centre results**

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**Introduction:** Aortic coarctation occurs in 5–8% of newborns with congenital heart disease. Surgical treatment is an obligatory standard of clinical management in children with primary coarctation under 6 months of age.

The aim of this study is to present our experience in interventional cardiology procedures for primary and secondary aortic coarctation in neonates and infants.

**Methods:** 51 pts aged from 6 days to 12 months required balloon angioplasty, while in 6 pts stent implantation was done.

Primary coarctation of aorta appeared in 18 pts (10 newborns, 1 premature-700 g) aged from 6 to 189 days (mean 49).

Interventions were performed as an emergency management in pts not qualified for surgical techniques due to severe condition or additional pathology (critical aortic valve stenosis, multi-organ insufficiency, pulmonary hypertension or severe infection).

Recurrent or residual coarctation occurred in 33 pts, aged from 1.5 to 12 months (mean 168 days), coexisting usually with other complex heart defects (TGA+VSD+CoA, Taussig-Bing+CoA, VSD+CoA, IAA+VSD).

**Results:** In all pts interventional procedures were used efficiently. The mean pressure gradient decreased from 35.7 to 11.2 mmHg

in pts with primary CoA and from 37.8 to 8.6 mmHg in pts with restenosis (evaluated by cardiac catheterization).

Four newborns with additional critical aortic valve stenosis underwent simultaneous balloon valvuloplasty with good result.

**Conclusions:** Stent implantation was successfully done in 4 pts with primary, tubular stenosis of aortic isthmus and concomitant hypoplastic aortic arch and in 2 pts due to recoarctation.

**Conclusions:** Emergency percutaneous interventions in neonates and infants under 6 months of age with primary CoA are worth to consider.

#### P-264

##### **Percutaneous stent implantation in right ventricle outflow tract obstruction – single centre experience**

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**Introduction:** In patients with critical right ventricle outflow tract obstruction (RVOTO) and hypoplastic pulmonary arteries stent implantation can be an alternative to palliative surgical intervention, especially when anatomical conditions restrict surgical treatment. The aim of this study is to present our experience in interventional cardiology procedures for right ventricle outflow tract obstruction.

**Methods:** We performed percutaneous interventions in a group of 5 patients with critical RVOTO aged 7 days to 118 months. 3 of them were diagnosed with ToF + MAPCA'S + hypoplastic pulmonary arteries, 1 with DORV + VSD + PS and 1 with DORV + VSD + ATR. AP + MAPCA'S. 3 patients were referred to us for primary intervention, the remaining 2 received previous surgical treatment (Blalock-Tausig shunt, opening and reconstruction of RVOT). We performed 13 cardiac catheterizations with 8 stent implantations and 10 balloon angioplasties. In 3 patients we attempted balloon dilatation of RVOTO and MPA prior to stent implantation. 3 patients required implantation of additional stent later on, with further 7 angioplasties to redilate previously implanted stent(s) or to relief stenosis due to overgrowth within or proximal to stent(s).

**Results:** We implanted successfully 4 stents as a first step treatment. In 1 patient stent embolized to the right ventricle immediately after balloon withdrawal and had to be removed surgically. 1 patient underwent successful stent implantation with concomitant balloon dilatation and awaits next step treatment under ambulatory care. In remaining 3 patients Mc Goon ratio increased from 1,28 (range 0,78 to 1,5) to 1,37 (1,28 to 1,5) so as Nakata index from 65,3 mm<sup>2</sup>/m<sup>2</sup> (27,7 to 81,4) to 86,7 mm<sup>2</sup>/m<sup>2</sup> (84,9 to 94,5).

**Conclusions:** In presented small group of patients with critical RVOTO percutaneous interventions allowed for reconstruction of right ventricle outflow tract. However despite increase in McGoan ratio and Nakata index patients still remain poor candidates for total surgical correction.

#### P-265

##### **Extension of indications for percutaneous pulmonary valve implantation in native right ventricle outflow tract : should all patients be considered?**

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**Introduction:** Patient selection for percutaneous pulmonary valve insertion (PPVI) is widely accepted, being limited to patients having a right ventricle to pulmonary artery conduit. Little data has been reported regarding PPVI on patients having a native right ventricle outflow tract (RVOT).

We present our data regarding PPVI in native RVOT and discuss the specific requirements to make this technique safe and durable.

**Patients and methods:** We review patients included over the last 18 months in the prospective study (REVALV) for patients undergoing intervention for RVOT dysfunction. Only valved stent on native RVOT group is analyzed here. Eleven patients were included. We performed MRI, balloon calibration and angiography of the RVOT to all patients in order to define the RVOT morphology and to establish a personalized technique for each patient in order to implant a valved stent on the native RVOT. All patients undergoing valved stent implantation are previously pre-stented with a bare metal stent according to present recommendations.

**Results:** Initial dimensions for these patients were on the upper limit for the established criteria. Two had a diameter above 24 mm. In one case, the "Russian dolls" technique was used (one bare-stent inside another and valved stent inside both of them, in order to reduce diameter). For the other patient, "branch jailing" technique was used (left pulmonary branch was stented down to the pulmonary trunk in order to have an appropriate diameter for valved-stenting). Pulmonary valve was placed successfully in all cases. All but one had been pre-stented at same procedure than valvulation. Of those, one freshly implanted bare metal stent dislodged to the right pulmonary artery. Two extra bare metal stents were implanted in order to cover the branch to the trunk, and finally valved stent was placed with no further problems.

**Conclusions:** Percutaneous pulmonary valve implantation can be performed on patients having native RVOT with success. Pre-stenting should be performed in a previous intervention in order to ensure stabilization of the bare metal stent and to avoid dislodgements. MRI, angiography and balloon calibration are not discriminating criteria for discarding candidates if personalized techniques are established for each patient.

#### P-266

##### **Sutureless anastomosis with covered stents during hybrid surgery for rescue of hypoplastic – distal intrapulmonary arteries**

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**Background:** Management of hypoplastic and/or deep intrapulmonary arteries in adolescents with pulmonary atresia can be very challenging. We report on the use of interventional techniques during hybrid procedures to make sutureless connections between grafts and diminutive arteries, which are expandable if required.

**Methods:** Prospective study. Sternotomy; the diminutive artery is identified; stay suture & vascular clip is put at puncture site to mark entry point; the artery is punctured and a wire is advanced in the pulmonary artery; sheath is advanced into pulmonary artery; covered stent is mounted on balloon, vascular graft is slit around shaft; covered stent is deployed sitting across entry point artery, balloon left inflated; graft is slit over proximal end of stent, and tightened distally with a vascular clip; balloon is exchanged for bigger balloon : the proximal stent overlapped by the graft is then dilated into the graft until tight fit; stay suture is now fixed in distal graft to avoid dehiscence; balloon and wire are



withdrawn, vascular clamp across the graft to allow “dry” proximal anastomosis of graft as required.

**Results:** 2 patients, 3 anastomosis:

Patient 1: 13y; left PA thrombosed in infancy, diminutive 2 mm but patent beyond the hilus (retrograde wedge); procedure as above: 0.014” wire; 4/19 mm Jostent Graftmaster to connect the vessel; stent to 5 mm stretch Gore-Tex graft with 6 mm balloon. During follow-up the graft and stent were further dilated up to 6 mm.

Patient 2: 15 years, bilateral duct with disconnected pulmonary arteries; 2 neonatal modified Blalock-Taussig shunts; right shunt diminutive but left shunt thrombosed; patency of small left PA well beyond hilus. Procedure as above: 0.035” wire in PA; 11F sheath, bilateral Covered CP stent on 12 mm balloon sequentially opened, connected to 14 mm Gore-Tex graft with 16 mm balloon. All anastomoses in time dilatable up to 18 mm.

In all procedures perfect hemostasis was obtained with good clinical result.

**Conclusion:** The sutureless connection with a covered stent allows successful rescue surgery on diminutive distal pulmonary arteries. This type of connection is further dilatable if required.

#### P-267

##### **Relief of Left Pulmonary Artery stenosis with stent implantation in children with Single Ventricle Malformation prior to Fontan**

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**Introduction:** Patients with single – ventricle Fontan physiology need an energy-efficient circulatory system to minimize the work load on the single ventricle. Pulmonary artery stenosis is a common lesion that limits the efficacy of the Fontan operation.

The Norwood Procedure (NP) as others surgeries with aortic reconstruction resulting in a large aorta, limits left pulmonary artery size based on their anatomic proximity.

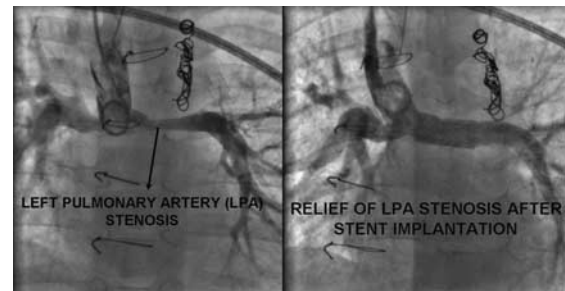
Our objectives are to evaluate the safety and efficacy of stent angioplasty of stenotic left pulmonary artery (LPA) in these patients, and to determine the mid-term outcome following these procedures.

**Methods:** Retrospective review of medical records; study period from January 2006 to December 2010. Inclusion of all pediatric patients with single ventricle malformation and bidirectional Glenn stage in which LPA stenosis was treated with stents. Initial, follow-up catheterization and surgical data were reviewed.

**Results:** During the study period 38 stents were implanted in the LPA in 37 patients. Mean (SD) age was 3.9 years (1.8). Mean (SD) weight was 15.2 Kg (1.3). 31 patients (83%) underwent NP as stage I palliation. The mean  $\pm$  SD size of the pulmonary artery stenosis was  $3.9 \pm 1.33$  mm before and  $8.3 \pm 1.6$  mm ( $p < 0.001$ ) after stent implantation. Redilatable (not premounted) stents were used in all but 3 patients. There were two procedure related complications: 1 stent migration and 1 left main bronchus compression in a 5 kg patient resolved by intrabronchial covered stent implantation. 24 of 37 patients had follow-up catheterizations after a mean time interval of 14 (0.2–48) months. Mean  $\pm$  SD pulmonary artery pressure decreased from  $11.9 \pm 3.1$  mmHg to  $11.1 \pm 2.7$  mmHg ( $p = 0.16$ ). In 4 patients (20%) in stent stenosis by significant neointimal proliferation was diagnosed, requiring balloon redilation. Fontan procedure was performed in 14 patients (37%) during the follow-up period with no technical inconveniences.

**Conclusions:** Stent implantation to treat left pulmonary artery stenosis in single ventricle patients is effective and can be

performed safely. The frequent compressive etiology of stenosis after NP makes this technique especially valuable. The use of redilatable stents permits long term treatment to optimize pulmonary hemodynamics. During total cavopulmonary connection previously implanted stents do not interfere with the surgery technique.



#### P-268

##### **Stent coarctation repair in older children and adults. Single group experience and long term follow up**

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**Introduction:** Transcatheter approach is emerging as the elective treatment for aortic coarctation in older children and adults. We describe our experience emphasizing on the initial results, technical details and long term follow-up.

**Methods:** Retrospective study in older children and adults with aortic coarctation treated by a single group for a period of 13 years.

**Results:** From 1997 to 2010, 53 patients [age: 19 (min. 8.7) years, weigh  $59.5 \pm 16.1$  Kg] were treated for native aortic coarctation (18) and recoarctation (35). Elective indications included hypertension, associated aortic regurgitation, peak systolic gradient  $>20$  mmHg and/or severe stenosis on CT/MRI. In 2 cases patients were in extreme low cardiac output. Technique: 54 stents were implanted in 53 patients. Balloon diameter was selected similar to diaphragmatic aorta without oversizing the arch or isthmus. BIB balloon's used in 38. Stent types: Palmaz iliac: 9, CP: 31 (covered 7), Palmaz Genesis XD: 6, EV3 MAX: 6, Atrium stent: 2. Rapid pacing during deployment was used in the last 22 cases.

**Immediate results:** Lesion diameter significantly increased from  $6.9 \pm 2.4$  mm to  $13.5 \pm 3.3$  mm and systolic gradient decreased from  $35.7 \pm 18.3$  to  $6.4 \pm 6.1$  mmHg. We had no mortality or mayor complications. Stent displacement happened in 2 initial cases. Follow up: mean  $4.4 \pm 3.1$  years in 44 patients: Two unrelated late deaths, one awaiting heart transplant. The only major complication: 1 severe aneurysm successfully dealt with percutaneous endoprosthesis, years after. Arterial access point injury was surgically solved in 3 patients. 6/16 patients went off hypertensive treatment. Peak echo gradient dropped from  $47.2 \pm 13.0$  to  $21.8 \pm 12.6$  mmHg. 8 fractures were detected: 3 after redilation. Eight patients required stent redilation for hypertension and/or residual gradient without significant complications: Gradient decreased from  $24.6 \pm 10.1$  to  $10.1 \pm 7$  mmHg and diameter increased from  $9.8 \pm 2.7$  to  $12.5 \pm 2.7$  mm.

**Conclusions:** Stenting aortic coarctation is a safe and effective technique in short and long term follow up. BIB balloons, rapid



acing and covered stents have improved the initial outcome, lessening complication rate. Restenosis can be redilated with good results. Stent implantation should be the elective treatment for aortic coarctation in the referred age group.

#### P-269

##### **Late complications after Interventional Closure of Atrial Septal Defect with the Solysafe® Septal Occluder**

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Prospective mono-center study for interventional closure of atrial septum defect, secundum type (ASD) by Solysafe® Septal Occluder (SSO) was performed in our institution, with one year follow-up. Patients were re-examined after implantation of a SSO, because of device fracture (DF) diagnosed in one patient.

**Objectives:** To evaluate long term results of ASD closure by SSO and potential complications, such as: device deformation (DD), device fracture (DF), wire embolization (WE), device thrombosis (DT), and others.

**Methods:** Extended follow-up examination was performed in all patients after SSO, including transthoracic echocardiography (TTE), standardized fluoroscopy (sF) and in pts with complications transesophageal echocardiography (TEE) as well.

**Results:** Between 09/2006 and 04/2008 57 pts have undergone SSO due to ASD. Median age was 15.9 years (4.5–53). Complete follow up was available in 49 pts (85.96%). Only one patient was symptomatic. Median follow-up was 3.2 years (2.25–3.7). Patients were divided in three groups: Group A- 12 pts with small SSO. (15 mm), Group B- 25 pts with medium size SSO. (20 and 25 mm) and Group C- 12 pts large SSO. (30 and 35 mm). Complications were diagnosed in 15/49 (30.6%) pts. Patients from Group A have not had any complication until now. In Group B 2/25 (8%) pts had DD and 1/25 (4%) pts had DF. In Group C 3/12 (25%) had DD and 9/12 (75%) pts had DF, from whom WE was disclosed in 3 (left atrium, right ventricle and right pulmonary artery) and DT in one. There is a significant correlation between size of SSO and the complication rate. All complications, except DT were diagnosed only by sF, while TTE was normal. Massive DT was diagnosed by TTE and confirmed by TEE. Surgical removal of SSO was performed in 3 pts with DF and DT. Unfortunately, one patient died after surgery due to massive pulmonary embolism, but there is no hard evidence to blame it to the device.

**Conclusions:** The incidence of complications after ASD closure by SSO is extremely high, particularly in pts with large SSO. sF is imperative for accurate diagnosis of DF and WE. Close monitoring is necessary for all pts with SSO.

#### P-270

##### **Percutaneous pulmonary valve implantation (Melody® valve) in the failing pulmonary valved Contegra® conduit in children and young adults**

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**Introduction:** Since CE market approval in 2006 Melody® transcatheter pulmonary valve implantation has been an interventional treatment option for the dysfunctional right ventricular outflow tract conduit. The aim of this new procedure is to restore

pulmonary valve competence without the need of open chest operation and therefore potentially reduce the number of open-chest interventions over a patients lifetime.

We present our first experience in percutaneous Melody® valve implantation in the failing pulmonary valved Contegra® conduit in children and young adults.

**Methods:** Between May 2010 and December 2010 we implanted 10 Melody® valves in 10 patients with a failing pulmonary valved Contegra® conduit. Patient age ranged from 9 to 23 years (mean 14.1 years) and weight from 30.7 kg to 79.2 kg (mean 48.6 kg). Contegra® conduit size ranged from 12 mm to 22 mm (mean 17.6 mm) with a conduit lifespan from 5 to 11 years (mean 9 years). All Contegra® conduits were pre-stented with Andra® XL and/or Cheatham platinum (CP) stents. The number of stents required to achieve a rigid tube without any re-coil as a Melody® valve landing zone in the right ventricular outflow tract ranged from 1 to 5 stents (mean 2.2 stents). All calcified Contegra® conduits were pre-stented with at least 1 covered CP stent. The Melody® valve was implanted in 7 patients on a 22 mm delivery system, in 2 patients on a 20 mm delivery system and in 1 patient on a 18 mm delivery system.

**Results:** All Melody® valves were successful implanted with no procedural complications. Residual gradients between RV and PA ranged from 0 mmHg to 15 mmHg (mean 7.8 mmHg). Pulmonary angiograms after valve implantation demonstrated a functional Melody® valve with no pulmonary insufficiency in all patients.

**Conclusion:** In our limited experience percutaneous implantation of the Melody® valve in the failing pulmonary valved Contegra® conduit can be performed safely. Whether these encouraging short-term results will be stable in the long term needs to be investigated.

#### P-271

##### **Detachable coils versus Amplatzer Duct Occluder devices in transcatheter treatment of small-to-medium sized Patent Ductus Arteriosus: an analysis of costs and results**

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**Objectives:** To review our experience in transcatheter closure of small-to-medium sized patent ductus arteriosus (PDA) with detachable coils and Amplatzer Duct Occluder (ADO) devices, and to compare the results with the two devices.

**Methods:** Between 1994 and 2010, 127 patients (pts) (median age 4,8 years, median weight 20 kgs) with a PDA size between 2 and 3,5 mm (median 2,4) underwent transcatheter PDA closure in our Institution. Procedural results were compared considering the type of device used in the first attempt; follow-up (FU) results were compared considering the type of device implanted. Costs were calculated considering the type and number of devices used (even if not implanted) for the procedure, excluding the fixed costs of the cardiac catheterization. A p value <0,05 was considered statistically significant.

**Results:** Procedural Results: there were 95 coil-procedures and 32 ADO-procedures. The mean fluoroscopy time was  $14,3 \pm 11$  and  $12,4 \pm 4,7$  minutes for coils and ADO, respectively (p = n.s.). There were 3 (3,2%) coil embolization, no embolizations for ADO (p = n.s.). Multiple attempts were necessary in 5 (5,3%) and in 1 (3%) pts (p = n.s.), and the procedure was successful in 93 (98%) and 32 (100%) pts (p = n.s.) for coils and ADO, respectively. The 2 pts with failure of coil implantation were treated with ADO devices in the same procedure. FU Results: residual shunt at discharge was present in 8/93 (9%) coil-pts and

in 2/34 (6%) ADO-pts ( $p = n.s.$ ). At the time of last FU visit, residual shunt was present in 2/93 (2,2%) coil-pts and 0/34 (0%) ADO-pts ( $p = n.s.$ ). One pt with RS was treated with an additional coil-procedure. Costs: the mean cost of the procedure was significantly lower for coils than for ADO. ( $538,5 \pm 415$  vs  $2436 \pm 294,2$  €,  $p < 0,001$ ).

**Conclusions:** In our experience of transcatheter closure of small-to-medium sized PDA, detachable coils compare favourably with ADO devices in terms of procedural and follow-up results. Given the great difference in costs, coils are a very cost-effective treatment option.

#### P-272

##### **Two Melodies in Concert: catheter-interventional double valve replacement**

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**Introduction:** Percutaneous pulmonary valve implantation (PPVI) has been established as a valuable treatment option for elder children and adolescents with conduit failure in the right ventricular outflow tract (RVOT). Transcatheter valve implantation in the tricuspid position is restricted to single case reports.

**Patient and methods:** A 26 year-old male initially diagnosed with tetralogy of fallot and hypoplastic pulmonary arteries hitherto underwent a total of 5 open chest procedures (BT-shunt, corrective surgery with transannular patch, pulmonary homograft implantation and tricuspid annuloplasty, tricuspid valve replacement with 27 mm bioprosthesis, pulmonary homograft exchange (26mm). He now presented with a degenerated pulmonary homograft (PR) and tricuspid bioprosthesis (TS; TR) in NYHA functional class III with severe signs of right heart failure, grossly dilated right atrium and ventricle and severely reduced RV function. Echo showed a 2cm IVC width with pendulum flow, tricuspid E wave of 2.4 m/s and a mean pressure gradient of 11 mmHg.

After a full hemodynamic study and coronary angiograms, the RVOT was pre-stented using a Plamaz P 4014 on a 20 mm BiB-Ballon followed by a Melody stent implantation on a 22 mm delivery system. The calcified tricuspid bioprosthesis was pre-dilated with a 20 mm Mullins high pressure balloon followed by a second Melody stent implantation in the tricuspid position with a 22 mm delivery system.

**Results:** Immediately after the second Melody valve implantation the RA pressure dropped from 22 to 15 mmHg, SVC oxygen saturation increased from 57 to 70% reflecting an improved cardiac index. Echocardiographic assessment at immediate and at 6 months follow-up showed a reduction of IVC width from 20 to 14 mm with no significant gradient across the Melody valve in tricuspid ( $v_{max}$  1.5 m/s) or pulmonary position ( $v_{max}$  1.4 m/s) and trivial (tricuspid) and no (pulmonary) detectable regurgitation.

**Conclusion:** In carefully selected patients not only the failing conduit in pulmonary position but also a degenerated and calcified bioprosthesis in the tricuspid position can be successfully treated by percutaneous valve implantation. Nearly 50 years after the first surgical double valve replacement, we report on the first catheter-interventional double valve replacement.

#### P-273

##### **Our experience in comparing Amplatzer duct occluders I and II**

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**Background:** The Amplatzer Duct Occluder (AGA Corporation, MN) is a self-expanding nitinol device for occlusion of patent ductus arteriosus (PDA). This device has revolutionised the treatment of PDA because of the ease of placement and higher rates of occlusion for all sizes and types of PDAs. Clinical results using the ADO demonstrated low rates of dislodgement and extremely high occlusion rates with a single device and procedure.

**Objective:** To compare our experience of duct occlusion using the amplatzer devices ADO1 and ADO2, document the immediate/early closure rate, complications and device behavior during implantation.

**Method:** Retrospective cohort study from July 2008 to December 2010. Review and evaluation of data of all patients on whom we attempted closure of PDA using the amplatzer device.

Demographics	ADO1 (median; range)	ADO2 (median; range)
Age in months	32; 11–180	16; 6–130
Weight in Kg	14; 8–61	9.5; 5.3–39
Narrowest ductal diameter in mm	2.5; 1.8–5	2.5; 1.5–4
Diameter of aortic ampulla in mm	8; 5–12	7.8; 4–13
Length in mm	5; 4–7	7; 5–15
Procedure time	39; 22–75	48; 22–104
Fluoroscopy time	5.3; 3.8–13	6.5; 2.8–20
Ductal morphology <sup>1</sup>	A(13%)B(60%) C(27%)	A(36%)B(20%) C(35%)D(2%)E(6%)

**Results:** Total 85 (ADO1–36, ADO2–49) ADO1 devices were 2–6/4, 23–8/6, 11–10/8, ADO2 devices were 24–4/4, 11–3/4, 9–5/4, 2–6/4, 2–4/6 and 1–5/6. In our unit, the approach is always from venous side. Follow-up echocardiography was done on D1 and 4–6 weeks post implantation. In ADO1 all except one implantation were technically successful with immediate complete angiographic closure in 34, trivial contrast flow in one patient and mild flow in one patient (that device embolised on the same day, removed surgically). In ADO2 immediate complete angiographic closure seen in 34, trivial contrast flow in 12 patients and mild flow in three patients of which two embolised. One was retrieved and closed with ADO1, the other was taken out surgically. There were no procedure-related complications.

**Conclusion:** Amplatzer duct occluders are safe and effective for PDA occlusion. In our experience ADO1 has a high early closure rate with lesser complications. ADO2 are more suitable for small children and longer ducts.

#### P-274

##### **Safety and Efficacy of Catheter Interventions in Premature Infants Under 2 kg**

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Premature infants with critical congenital heart disease pose difficult therapeutic problems: Surgery is associated with 1.5–3.0 times higher mortality and prolonged medical therapy to achieve weight gain does not improve survival. Therefore, we elected to perform early therapeutic or palliative catheter intervention. To assess safety and efficacy of this approach, we collected demographics, procedural parameters, results, complications and late

clinical outcomes of all premature infants <2 kg undergoing attempted catheter intervention.

Interventions were attempted in 16 patients and accomplished in all. Access was from jugular or carotid cut-down in patients <1.5 kg, otherwise percutaneously from femoral vessels. Fluoroscopy times were 2.3–35.8 min (median 14 min). Balloon atrioseptostomy was performed in 1 (weight 1.6 kg). 3 patients underwent pulmonary balloon valvuloplasty (weight 1.0–1.98 kg) with no re-intervention during follow-up. 4 patients underwent aortic balloon valvuloplasty (weight 1.1–1.9 kg): no re-intervention in 1, one patient moribund at the time of procedure died the following day. One patient required repeat balloon valvuloplasty after 2.7 months and Ross operation after 4 years. This patient suffered right lower limb ischemia after initial femoral access and required foot amputation. One patient underwent Ross operation for endocarditis unrelated to the procedure after 4 months. 4 patients underwent palliative stent implantation for aortic coarctation (weight 960–1230g) with stents 3–4 mm in diameter and 7–9 mm in length (fluoro time 6.2–10.8 min). One patient died from sepsis; 2 had elective end-to-end repair after 4 months; 1 underwent Norwood after 3 months. 3 patients underwent palliative stent placement for Tetralogy of Fallot (weight 970–1300 g) with stents 3.5–4 mm in diameter and 9–12 mm in length (fluoro time 10.8–17.2 min). Elective repair was performed in all after 6–10 weeks. One patient with HLHS underwent stent implantation into the atrial septum for severe atrial restriction (weight 1400g) but died 7 days after bilateral PA banding 2 weeks after atrial stent secondary to severe native aortic arch stenosis.

Catheter intervention offers relatively safe and effective treatment or palliation for premature infants under 2 kg but overall outcome in critical AS and HLHS leaves room for improvement.

#### P-275

##### Results of endovascular stenting of coarctation of the aorta in 189 patients – Polish registry

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**Objective:** To describe the combined experience and outcomes of stent implantation in patients with native (CoA) and post-operative (rCoA) coarctation of the aorta performed in four Polish cardiology centers.

**Methods:** Retrospective data collection was analyzed. Primary endpoints were peak systolic catheter gradient reduction, stented segment diameter increase. Early and late complications, changes in antihypertensive medication after stenting were recorded.

**Material:** Between 1997 and 2009, 189 pts with arterial hypertension underwent stent implantation (CoA– 142 pts, rCoA– 47 pts). For primary treatment 192 stents were used (52 Palmaz, 84 Cheatham–Platinum, 34 covered Cheatham–Platinum, 22 other). Median patients age was 15.5 yrs (5–57) – 74,6% were below 18 yrs, 10% above 40 yrs.

**Results:** There was significant improvement ( $p < 0.001$ ) in pre versus post stent coarctation diameters (5.45  $\pm$  2.69 mm (1–16, med.5) vs. (14.23  $\pm$  3.19 mm (5.2–22, med.14) and systolic gradient (42.98  $\pm$  16.32 mmHg (15–111, med.40) vs.

7.45  $\pm$  8.70 mmHg (0–37, med.4). Successful reduction in the post stent gradient (<20 mmHg) was achieved in 91,9% of primary procedures. Acute complications were encountered in 10/189 (5%) procedures (stent migration –6, stroke –1, transient arm neuropraxia –2, acute aorta dissection–1). During follow-up period 4.43  $\pm$  3.17 yrs (0.2–14, med.4) 55% of pts did not need antihypertensive treatment and all the others have better control of arterial hypertension on lower doses of medications. About 55% (103/189) of the procedures were followed up by CT/MRI/angiography and confirmed good post-procedural anatomy in the first year after stent implantation. Stents fracture (5) and neointimal hyperplasia (8) were confirmed in CT during later follow-up. Additional procedures were performed in 46/189 pts (24,33%) – stent redilation due to planned staged procedure, intimal hyperplasia and/or patients growth –34, covered stent implantation due to small aneurysms –5, aortic arch narrowing –2, stent fracture –5.

**Conclusions:** 1. Stent implantation in native and postoperative coarctation of the aorta has good acute, intermediate, and long-term outcome 2. Continuous follow-up of patients after stent treatment of aortic coarctation is required due to associated long-term morbidity related to aortic wall complications, systemic hypertension, recurrent obstruction and need for additional interventions.

#### P-276

##### Percutaneous Closure of “tunnel shaped” ventricular septal defect using the Amplatzer Vascular Plug II in pediatric patient

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**Introduction:** Surgical closure is still considered the gold standard treatment of Ventricular Septal Defects (VSDs). Nevertheless some defects, such as apical muscular VSDs, are poorly accessible for traditional surgical approach. With introduction of Amplatzer VSD occluder devices (AGA Medical Corp., Golden Valley, MN, USA), VSDs transcatheter occlusion appears to be a valid alternative to cardiac surgery. However, tunnel-shaped VSDs are difficult and challenging to close with Amplatzer VSD occluder device (AGA Medical Corp., Golden Valley, MN, USA) due to the not appropriate device morphology. We report the first successful closure of a large tunnel-shaped apical muscular VSD with an Amplatzer vascular plug II (AVP II- AGA Medical Corp., Golden Valley, MN, USA) in a 27 month-old female patient.

**Methods:** A 27 month-old, 7.3 kg baby-girl was accepted at our institution for multiple VSDs. Echocardiographic evaluation showed a large perimembranous VSD (p-VSD) and an apical muscular VSD (m-VSD). M-VSD closure was considered to be challenging with traditional surgery. We decided to close m-VSD percutaneously. Right and left heart catheterization were performed and the apical m-VSD size was measured after left ventricular (LV) angiography. The m-VSD appeared to be “tunnel shaped”, similar to a cylindrical vascular structure, therefore, we selected for closure an AVP II (AGA Medical Corp., Golden Valley, MN, USA), a cylindrical self expandable occlusion device which is usually used for transcatheter embolization of unwanted blood vessel. The patient underwent p-VSDs surgical closure the day after.



**Results:** AVP II (AGA Medical Corp., Golden Valley, MN, USA) was correctly deployed and immediate post-procedure LV angiography showed trivial residual shunt. The patient remains symptom free with optimal combined procedure result, at 6 months follow-up.

**Conclusion:** A combined surgical and percutaneous procedure can be an easier and safer alternative to surgical closure only in selected VSDs cases. AVP II appears user- and patient-friendly device also in positions and in conditions for which it has not been designed ensuring an excellent procedural result.

#### P-277

##### **Results of transcatheter balloon aortic valvuloplasty in 59 neonates – one center experience**

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**Objectives:** The aim of this study was to evaluate the results of transcatheter balloon aortic valvuloplasty (BAV) performed in neonates with critical aortic valve stenosis (AS).

**Background:** Aortic valve function and reinterventions after BAV in neonates are not well characterized.

**Methods:** From 90 neonates with critical AS who underwent BAV in our center in period 1996–2006, 59 pts are in continuous follow-up. Retrospective and prospective data collection of these pts was analyzed. All procedures were performed in general anesthesia through carotid artery cut down or femoral artery puncture at the median age 6,5 days (1–28). Diameter of balloon did not exceed the aortic valve ring diameter in any case.

**Results:** There was significant systolic gradient decrease ( $p < 0.001$ ) pre versus post BAV. ( $58.49 \pm 24.82$  mmHg (25–150, med 55) vs.  $15.81 \pm 11.03$  mm Hg (0–45, med.15). Acute post-dilation aortic regurgitation was severe in 3/59 pts (5%).

The median follow-up period was 8 years (3–16.5); cumulative follow-up 536 patient-years. During follow-up systolic gradient increased significantly to  $30.24 \pm 11.53$  mmHg (9–57, med. 31), mean systolic gradient to  $17.9 \pm 11.2$  (3–62, med.15). Group of pts with severe aortic regurgitation increased to 18/59 pts (30%).

During follow-up median z-score of aortic valve annulus increased from 0.4 (–2.4–2.84) to 1.02 (–1.16–3.79) ( $p < 0.01$ ). Ascending aorta diameter increased significantly (median z-score before 1.82 (–1.63–5.5), after 3.46 (1.15–8.29) ( $p < 0.01$ ).

Nine pts (15.3%) median age 8,75 yrs (0.33–13.08) had surgical intervention for aortic valve (aortic valve repair –2, aortic valve replacement –7), 3 pts additional balloon valvuloplasty. Freedom from aortic valve interventions was 47/59 pts (79,66%).

**Conclusions:** 1. Transcatheter aortic balloon valvuloplasty is effective for relief of congenital aortic valve stenosis in neonates. 2. Continuous follow-up of patients after balloon aortic valvuloplasty is required due to long-term hazards for surgical aortic valve reintervention and replacement.

#### P-278

##### **Catheter Rehabilitation of Occluded Aberrant Right Pulmonary Artery**

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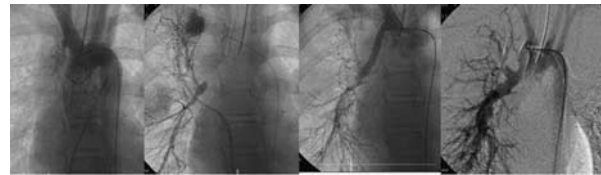
**Introduction:** A 10 year old asymptomatic girl was found to have right lung hypoplasia during routine chest X-ray for a viral

infection. CT chest documented an absent right pulmonary artery, but normal bronchial tree and a suggestion of pulmonary veins. Cardiac catheter documented a large left pulmonary artery and a diverticulum of Kommerell on the underside of a right innominate artery in the setting of a left arch. Pulmonary vein wedge angiography identified a hypoplastic disconnected right pulmonary artery system. We postulated that re-canalization of the occluded ductus arteriosus would re-establish flow to the RPA and promote growth, allowing for later surgical repair.

**Methods:** The occluded ductus arteriosus could be re-crossed with a straight 0.018 Terumo wire. This was exchanged for an 18 G perfusion catheter for test injections and for exchange for a 0.014" stiff coronary wire. The occluded duct was then stented to 5 mm diameter using 2 uncovered coronary stents. The procedure was uneventful.

**Results:** There was no change in exercise tolerance. Clinically and on ultrasound there was good flow through the stented duct. On repeat angiography the RPA system had grown significantly with an increase in diameter from 3 mm to 12 mm. The patient is now listed for surgical repair (interposition graft from MPA to RPA).

**Conclusions:** Unilateral lung hypoplasia, in the setting of a normal bronchial tree and absent pulmonary arterial supply should prompt the detailed search for a native pulmonary artery system. Catheter re-canalization should be attempted, in order to establish perfusion and growth.



#### P-279

##### **A Novel Technique for Stenting Pulmonary Artery and Conduit Bifurcation Stenoses**

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**Introduction:** Despite advances in endovascular stent materials and techniques, pulmonary artery bifurcation stenosis remains a challenging lesion. Congenital heart disease requiring a right ventricle to pulmonary artery conduit can present with either distal conduit or proximal bifurcation stenosis. These lesions are difficult to treat without surgery. Previous techniques include “kissing stents” or stenting of the major vessel, whilst straddling the smaller vessel. We describe a novel technique where a single stent is mounted on two angioplasty catheters. This stent is delivered using a large Mullins sheath on two guidewires, one in each of the bifurcating vessels.

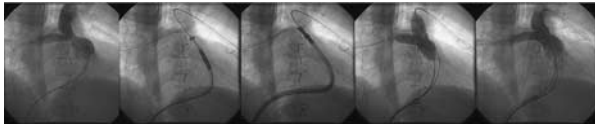
**Methods:** A superstiff guidewire was placed in the larger of the two bifurcating vessels. A wide bore Mullins sheath is advanced just proximal to the stenosis. A standard exchange wire is placed in the adjacent vessel using a 5 French right Judkins catheter. Both EV3 and CP stents were used because of limited foreshortening characteristics. The stent was crimped manually over the two angioplasty catheters. The catheters were advanced over respective guidewires through the Mullins sheath and into the branching vessels. Test angiograms through the side arm of the Mullins confirmed stent position before simultaneous balloon inflation to produce flaring of the stent. Nine patients (mean age 13.2 (6.7–23.4) years, mean weight 43.1 (23–69) kg) had a bifurcation stenosis treated using this technique between 2003



and 2010. Outcome measures were reduction of gradient, ratio of right to left ventricular pressures, procedural complications and need for re intervention.

**Results:** There were no significant complications during the procedure. The pressure gradient across the stenosis was reduced from 34 (23–52) mmHg to 11 (8–15) mmHg ( $p < 0.05$ ). Right ventricular to left ventricular pressure ratio decreased from 0.81 (0.55–0.95) to 0.42 (0.35–0.50) ( $p < 0.05$ ). No patients have required re-intervention or surgery over a median follow up of 2.6 (0.2–6.7) years.

**Conclusions:** This technique appears to be safe and effective for selected cases with bifurcation stenosis. It significantly reduces the risk of jailing side branches and the risk of intimal damage from using one oversized balloon. It should be considered a valuable adjunct in the catheter treatment of conduit stenosis.



#### P-280

##### The Three-dimensional Morphological Changes of tricuspid Valve in Children after Transcatheter Closure of Atrial Septal Defect

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**Background:** The closure of secundum type atrial septal defect (ASD) provides an decrease and normalization of originally volume overloaded right ventricular (RV) volume in ASD patients. However, three-dimensional (3D) morphological change of tricuspid valve (TV) remains unclear by this change of RV volume coming along with ASD transcatheter closure.

**Objectives:** The purpose of this study is to characterize the 3D morphology of TV in ASD patients comparing to those in normal subjects and to assess the 3D geometric change of TV by ASD closure, using real-time 3D echocardiography.

**Methods:** 11 children underwent ASD transcatheter closure with Amplatzer septal occluder (ASO) (age:  $8.3 \pm 3.6$ , Qp/Qs:  $2.4 \pm 0.6$ ). Their RV full-volume data of 3D echocardiography were recorded on hard disk by iE33 (Phillips Healthcare, Andover, MA) with X7-2 and X3-1 probe, before ASO, at one to 3 days and 6–12 months after ASO, respectively. The right ventricular dimension was measured with conventional 2D images. Tricuspid 3D morphology were analyzed and quantified by the off-line analyzing software: REALVIEW<sup>®</sup> (YD Ltd., Osaka, Japan) after obtaining 3D volume data by QLAB (Phillips) We measured annular area (AA) and circumference (AC), maximum tenting length (TeL), tenting volume (TeV), tenting area (TeA) at mid-systole, and compared those before and after ASO. These values were compared with those of age-matched normal controls (10 cases), after correction by body surface area.

**Results:** ASD patients before ASO had larger TeV and TeA than controls ( $0.81 \pm 0.4$  vs.  $0.54 \pm 0.3 \text{ cm}^3/\text{m}^2$ ,  $7 \pm 1.5$  vs.  $5.4 \pm 1.2 \text{ cm}^2/\text{m}^2$ ,  $p < 0.01$ ). Comparing values before and after ASD closure, AA, AC and TeA were significantly decreased at 6–12 months after ASO (AA:  $5.7 \pm 1.3$  vs.  $4.1 \pm 0.9 \text{ m}^2/\text{m}^2$ , TeA:  $7.0 \pm 1.5$  vs.  $5.1 \pm 1.1 \text{ cm}^2/\text{m}^2$ ,  $p < 0.05$ ). The other TeL and TeV were also decreased at long after ASO but statistically not significant.

**Conclusions:** Children with ASD demonstrate larger TeV and TeA than normal because of the enlargement of TV annulus and

leaflet. Comparing values before and after ASO, an decrease of all 3D morphometric parameters of TV, (especially AA, AC, and TeA) was presented after ASO as reduced RV volume. Since TeL and TeV shows no significant change, these changes of TV shape may improve coaptation of TV valve and reduce tricuspid regurgitation.

#### P-281

##### A twenty Year Experience of Arterial Switch Repair by Direct Anastomosis Retaining a Normal Position of the Pulmonary Bifurcation

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**Objectives:** The arterial switch operation (ASO) has become the procedure of choice for correction of transposition of the great arteries (TGA) since its introduction by Jatene in 1976. The Le Compte manoeuvre is the standard technique used to reconstruct the neopulmonary artery, but this has led to reports of a significant incidence of pulmonary artery stenosis (PAS). We have reviewed our experience with anatomic correction of TGA, with direct pulmonary artery anastomosis, avoiding the routine use of the Le Compte manoeuvre.

**Methods:** From October 1990 to March 2010, 101 patients underwent correction of TGA, using the ASO in our institution. There were 67 males (66.3%). Median age was 12 days (range 2–718). Sixty two patients (61.38%) were simple TGA with intact ventricular septum (IVS), and 39 patients (38.61%) were complex TGA. Follow up was 100% complete. Patients were assessed for PAS, aortic incompetence, coronary ischaemia and functional class.

**Results:** There were 8 early (7.92%) and 2 late (1.98%) deaths. Five patients underwent reoperations. Significant pulmonary artery stenosis has occurred in 5 of 91 surviving patients (5.49%). Mean pulmonary artery pressure gradient was 14.36 mmHg (range 0–42 mmHg). Aortic regurgitation was trivial in 27 patients, and mild to moderate in 9 in patients. Ninety eight patients (97%) were in functional class 1 with no symptoms or signs of ischaemia.

**Conclusion:** The arterial switch repair by direct anastomosis, retaining a normal position of the pulmonary bifurcation is feasible with an acceptable mortality and a low incidence of significant pulmonary artery.

#### P-282

##### Risk Factors in Neonatal Cardiac Surgery

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**Basics:** In recent years more researches of leading hospitals are devoted to analysis of different factors, which augment mortality risk in newborns with congenital heart disease. Risk factors differentiation helps to analyse outcome and quality of life after cardiac surgery in newborns.

**Material and methods:** From 2005 till 2010 we have treated 7558 children 1st year of life, incl. 1193 (16%) newborns (av. age 3,6 days, av. weight 2,9 kg). Congenital heart defects were the following: TGA, LHHS, coarctation of the Aorta, PA atresia, critical aortic or pulmonary valve stenosis, etc. As to the structure

of surgical interventions for the whole group, 54% (4065 operations) were made under artificial circulation (283 of which were operations for newborns, 40% were urgent, with survival rate of 86,9%) and 46% under closed methods.

**Results:** The statistic analysis of the factors influencing the outcomes of the surgical treatment ( $p < 0,01$ ) in conjunction with the expert estimate of anamnestic, clinical, laboratory results of the neonates study lead to the conclusions on the risk factors. In the neonates group these are: prematurity and low birth weight (339 children – 31,6%), hypotrophy (82 children – 7,6%), early neonatal period (468–39,2%), TORCH infections (56 children – 5,2%), concomitant somatic pathology and mechanical ventilation while entering hospital (216 children – 20,1%), multiple disemбриogenic stigmata and syndromic forms of CHD. (94 children – 8,8%). Severity of condition was characterized by the presence of the risk factors in each child (from 0 to 6). The increase in the quantity of risk factors lead to the increase ( $p < 0,05$ ) of the mortality risk. Analysis of the risk factors enabled to elaborate particular dynamic steps of clinical treatment for each group in order to improve the outcomes. As a result in a period from 2005 to 2010 the number of newborns significantly ( $p < 0,03$ ) increased and the survival rate after urgent operations improved to 94,2%, incl. premature and low-birth-weight children (av. weight 1,6 kg).

**Conclusions:** Optimization of preoperative care in children with CHD and risk factors exerts positive influence on quality of treatment and improves outcome for newborns and infants with CHD.

#### P-283

##### **Correlation of near-infrared spectroscopy (NIRS) with cardiac index in infants following surgery for congenital heart disease**

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**Objectives:** Determine the values of spectroscopic monitoring by INVOS system during the immediate postoperative. Compare the values obtained in the regional oxygen saturation (rSO<sub>2</sub>) in the flank (rSO<sub>2f</sub>), abdominal (rSO<sub>2a</sub>) and cerebral (rSO<sub>2c</sub>) with the central venous oxygen saturation (ScvO<sub>2</sub>) and cardiac output measured by transpulmonary thermodilution (TPTD). Assess the ability of INVOSR to detect the situation of low output.

**Material and Methods:** Prospective (February 2009 – June 2010) analytical study in children under 10 kilograms. Measurements were made at 1, 4, 8, 16 and 24 hours after surgery in PICU using INVOS 5100R, simultaneously ScvO<sub>2</sub>, stroke volume index (SVI) and cardiac index (CI) obtained by TPTD (PiCCO) were registered. Patient data are expressed as median and range. We used the Pearson test to determine the association between the variables obtained and the Mann-Whitney test for comparison of means.

**Results:** We included 17 patients, age 8 (3–34) months and weight 7.8 (3.8–10) kilograms. There was a significant correlation between ScvO<sub>2</sub> and right rSO<sub>2c</sub> ( $r = 0.54$ ), left rSO<sub>2c</sub> ( $r = 0.63$ ) and rSO<sub>2f</sub> ( $r = 0.54$ ) and a stronger correlation with the previously proposed formula =  $0,45x \text{ rSO}_{2c} + 0,45x \text{ rSO}_{2f}$  ( $r = 0.76$ ). SVI correlated with right rSO<sub>2c</sub> ( $r = 0.66$ ) and left rSO<sub>2c</sub> ( $r = 0.69$ ). In addition, we obtained a formula =  $-33 + 0.215x \text{ rSO}_{2f} + 0.626 \times \text{ rSO}_{2c}$  that correlates the flank and cerebral saturation with SVI ( $r = 0.72$ ). Comparing the measures that showed CI < 3.3 vs. CI > 3.3 ml/min/m<sup>2</sup> we obtained a significant difference  $p < 0.01$  for the right rSO<sub>2c</sub>, (65 vs. 73%), left rSO<sub>2c</sub> (64 vs. 73%), SVcO<sub>2</sub> (68 vs. 77%), proposed formula (66 vs. 72) and new IC formula (24 vs. 31 ml/m<sup>2</sup>).

**Conclusions:** We found an association between NIRS, ScvO<sub>2</sub> and TPTD. A cerebral and renal measurement together improves correlation slightly. Spectroscopy is useful to detect low output state. INVOS provides a useful monitoring the postoperative congenital heart disease in children under 10 kilograms.

#### P-284

##### **Cardiac output and intrathoracic volumetric parameters obtained by ultrasound dilution and transpulmonary thermodilution methods in a pediatric animal model**

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**Introduction:** Especially after cardiac surgery monitoring of cardiovascular function is essential in critically ill children. A novel minimal invasive indicator technique using ultrasound dilution (UDT) was recently introduced to measure cardiac output and intrathoracic blood volumes. Therefore, we compared cardiac output measurements and derived volumes using UDT (Transonic Systems Inc., NY, USA) with another indicator technique, the well-established transpulmonary thermodilution (TPTD) method (Pulsion medical systems, PiCCO plus, Germany) in a juvenile animal model.

**Methods:** Experiments were performed in 20 ventilated, anaesthetized piglets during isovolemic haemodilution. In each of the animals a 4-F Pulsiocath (Pulsion medical systems, PiCCO plus, Germany) catheter was surgically placed into the carotid artery and a central venous catheter introduced into the jugular vein. Piglets were randomly assigned into 4 groups and underwent four steps of isovolemic haemodilution by exchanging blood either with a colloid osmotic or crystalloid solution. Haemodynamically stable conditions provided ten minutes after each step of isovolemic haemodilution cardiac output and intrathoracic volumes were measured with TPTD followed by UDT. For each method, three injections were averaged. Cardiac output and intrathoracic volumes were compared by using bias and limits of agreement calculated as per the Bland-Altman approach and the linear regression analysis.

**Results:** A good agreement for measured cardiac output (Mean 1.98; range 1.1–2.9 l/min) with a percentage error of 17.3% with  $r = 0.92$ , mean bias = 0.28 L/min was observed. Global end-diastolic volume (GEDV) and intrathoracic blood volume (ITBV) measured by TPTD were almost 2 times larger than corresponding volumes quantified by UDT (ITBV(UDT) =  $0.58 \times \text{ITBV(TPTD)} + 27.1$ ; GEDV(UDT) =  $0.48 \times \text{GEDV(TPTD)} + 23.1$ ).

**Conclusion:** Cardiac output measured by UDT was found to be equivalent and hence interchangeable with TPTD in a paediatric animal model. The lack of agreement in the intrathoracic volumes recorded by two indicator methods could be due to either the different types of indicators (diffusible versus non-diffusible) or the underlying algorithm. Future studies are necessary to assign these results to critically ill paediatric patients and to determine the exact intrathoracic blood volume with reference techniques.

#### P-285

##### **Intravenous beta1-receptor blocking medication as a rescue treatment for severe postoperative myocardial diastolic dysfunction in infants**

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**Background:** Diastolic left ventricular myocardial dysfunction follows surgical repair of critical neonatal congenital heart defects despite successful initial treatment in some patients. Low cardiac output states present in many such patients lead to high levels of catecholamines with concomitant tachycardia and increased postoperative morbidity. We have used intravenous infusion of esmolol to lower heart rate and facilitate postoperative recovery in this setting.

**Case reports:** Case A: term neonate with critical aortic stenosis and endocardial fibroelastosis who underwent emergency balloon aortic valvuloplasty at 2 days of age followed by Ross-Konno procedure at 2 month. One month later he still required artificial ventilation and at cardiac catheterisation left ventricular end-diastolic pressure was 24 mmHg. Continuous infusion of esmolol was added to digoxin and milrinone reducing heart rate from 180 to 110 bpm. He was successfully extubated 3 days later.

Case B: premature neonate with hydrops and critical aortic stenosis who underwent surgical aortic valvuloplasty at 2 month of age. She remained ventilator dependent with evidence of normal left ventricular systolic function but persistent diastolic dysfunction. Continuous infusion of esmolol was added to enoximone reducing heart rate from 180 to 120 bpm. Ventilation was successfully weaned 4 days later.

Case C: preterm neonate who underwent anatomic repair of transposition of the great arteries complicated by apical left ventricular ischaemia and systolic and diastolic dysfunction. Systolic myocardial function gradually recovered but severe diastolic dysfunction persisted and the patient has been dependent on artificial ventilation for 10 weeks postoperatively. Intravenous esmolol infusion was added to enoximone and captopril treatment followed by reduction in heart rate from 150 to 115 bpm and weaning from positive pressure ventilation in 3 days.

**Conclusion:** Benefit of lowering heart rate in acute or chronic myocardial dysfunction has been well recognised in adult patients with favourable impact of beta-receptor blocking medication for several decades now. Our limited data show that the use of intravenous selective beta1-receptor blocker facilitates postoperative recovery in infants with persistent tachycardia and severe left ventricular diastolic dysfunction following cardiac surgery.

#### P-286

##### **Is oral enoximone a safe alternative to protracted intravenous medication in severe myocardial failure in children?**

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**Introduction:** Prolonged severe myocardial dysfunction presents difficult therapeutic challenges in the paediatric population. Intravenous phosphodiesterase 3 inhibitors have proven to be a valuable and successful therapy in paediatric patients with acute or chronic myocardial dysfunction over the last two decades. Unfortunately there is no specific oral preparation available. Continuous intravenous administration is associated with risk of infection and considerable patient discomfort. Weaning from intravenous medication to more commonly used oral Angiotensin-Converting Enzyme (ACE) inhibitors and beta-blockers can be challenging and poorly tolerated. We examined our experience with using the intravenous preparation of enoximone as an oral medication in this setting.

**Methods:** We reviewed retrospectively hospital records of 23 patients receiving oral enoximone in a single tertiary paediatric cardiac centre between November 2005 and December 2010.

**Results:** Patient age at start of oral enoximone was 1–173 months (median 8 months). Three patients (15%) had left ventricular dysfunction due to myocarditis or cardiomyopathy. Twenty patients (85%) had myocardial dysfunction complicating congenital heart disease, 19 (90%) following cardiac surgery. Of this latter group 9 (45%) had left ventricular dysfunction, 8 (40%) had right ventricular dysfunction and 3 (15%) had biventricular failure.

Indications for oral enoximone were inability to wean from intravenous milrinone infusion and/or intolerance of ACE inhibitor therapy. All patients received oral enoximone at 1 mg/kg 3 times/day.

Enoximone was well tolerated at this dose without adverse haemodynamic effect. Due to alkaline nature of this solution there were 2 (8%) patients with blood stained gastric content aspirates when enoximone was administered without milk, which subsequently resolved when given with milk. No other adverse effects were encountered and the families tolerated well the inconvenience of using an intravenous preparation with the need for refrigerated storage. Results of outcome data analysis will be presented.

**Conclusions:** Oral enoximone is a safe alternative to protracted intravenous treatment of severe myocardial failure in children. Based on our experience it is a well tolerated and promising alternative when ACE inhibitors and beta-blockers are not tolerated. The development of a licensed oral preparation would be welcomed.

#### P-287

##### **Age impacts on outcomes of children on mechanical ventricular support**

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The aims of this study are to report the first largest French experience with ventricular assist device (VAD) in children and assess the influence of age on outcomes.

**Material and methods:** From 2005 to 2010, 16 patients aged <18 years, needed pulsatile ventricular mechanical support at Lyon University Medical Center. Their clinical data, echocardiographic records and outcomes were reviewed.

**Results:** Seven females and 9 males, aged 0.3 to 16 years (med 2.3), were implanted with pulsatile VAD (<2years: 8 and >2years: 8): 6 with left ventricular support and 10 biventricular, for either cardiogenic shock with cardiac arrest in 4, or uncontrolled low cardiac output in 12. Underlying cardiac diseases included: 14 dilated cardiomyopathies, 1 acute myocarditis and 1 post-ischemic. Median hospital stay prior to implantation was 13days. Seven of 8 patients <2years (87.5%) and 50% of those >2years were on mechanical ventilation prior to assistance (p = 0.10). Duration of support was 6 to 125 days (med 37). Eleven patients were extubated while on support (68.7%). Five infants, all <18 months of age, experienced stroke due to cerebral embolia (30%), 3 had an hemorrhagic complication. Four patients died on support (25%), at 8th, 23rd, 60th and 108th day, from sepsis, multivisceral embolia, hemorrhage and canula rupture respectively, from those 3 ranged in the youngest group. Two patients were successfully weaned off support at 19th and 37th day (12.5%), 10 underwent heart transplantation (62.5%).

**Conclusion:** Although the thrombo-embolic risk and mortality on support are significantly higher in patients less than 2 years of age,



our experience of pulsatile VAD in children shows overall survival rates of 75%.

#### P-288

##### **Beyond Adenosine: Use of Dexmedetomidine in a New Approach for the Termination of Reentrant Supraventricular Tachycardia**

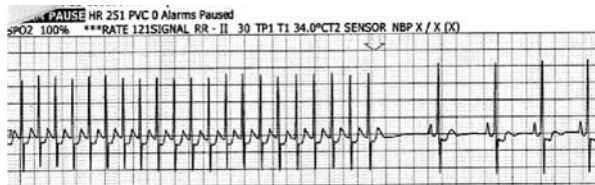
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**Introduction:** Dexmedetomidine an alpha-2 adrenoreceptor agonist has been shown to have potential novel anti-arrhythmic properties. In a recent case-series it was successfully used for the treatment of junctional and atrial tachyarrhythmias for conversion to sinus rhythm (SR) or heart rate control. Adenosine the typical agent for acute cardioversion of reentrant-supraventricular tachycardia (SVT) it's not without limitations. These include ultra-short duration, making it less than ideal with multiple paroxysmal episodes, unpredictable duration of asystole and feeling of impending death, atrial and ventricular fibrillation, and not infrequently ineffectiveness. We present an original approach for the acute cardioversion of reentrant-SVT using dexmedetomidine.

**Methods:** Retrospective review of 19 patients admitted to the Cardiac ICU and treated with dexmedetomidine.

**Results:** Median age was 10 days (1–827). Ten patients (53%) were postoperative after congenital cardiac surgery. Fourteen (74%) were on baseline antiarrhythmic agents (amiodarone, propranolol, digoxin) and 3 had history of WPW syndrome. Three patients (16%) had received 12 (9–2–1) doses of adenosine prior to dexmedetomidine with successful cardioversion in 8/12 doses (67%). Initial SVT rate was  $241 \pm 22$  bpm. After  $0.7 \pm 0.3$  mcg/kg of dexmedetomidine given at a bolus rate of 1.0 mcg/kg/min (0.3–6.0), cardioversion was achieved in all patients (100%) (Fig). Time to SR was 30 seconds (15–120). Three (16%) had a total of 5 recurrent episodes of reentrant-SVT at  $3.2 \pm 0.9$  hrs apart and were cardioverted with additional dexmedetomidine. None developed prolonged sinus pause, asystole or other arrhythmias. The median sinus pause duration was 0.7 seconds (0.4–1.1). Transient (<3 minutes) hypertension  $28 \pm 5\%$ , was observed when dexmedetomidine rate was more than 0.7 mcg/kg/min. One episode of hypotension resolved with a one low-dose phenylephrine bolus. Mild to moderate sedation was seen in all patients lasting for 35 minutes (15–60).

**Conclusions:** Dexmedetomidine appears to be a safe and effective agent for the acute cardioversion of reentrant-SVT.



#### P-289

##### **Advanced medical simulation serving pediatric cardiology: how can we increase the quality of our management without compromising patient safety?**

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**Introduction:** Management of pediatric cardiac patients involves unique and complex physiology requiring an adequate knowledge and training of a multidisciplinary teamwork. Medical simulation has showed to be a good method of teaching clinical knowledge and procedural skills which can enhance patient safety. The aim of the study is to describe the initial experience of medical simulation in training pediatricians in cardiac critical situations.

**Methods:** We planned a course to train neonatologists, pediatric intensivists, pediatric residents and cardiology residents using real scenarios with high-fidelity manikins. Debriefing followed each scenario, focusing on key points of the management of cardiac critical events including basic concepts on echocardiography. Postparticipation questionnaires were used to know the effects on the participants.

**Results:** A total of 20 providers participated in the course. All participants scored the usefulness of the program and scenarios as 4 of 5 or higher (5 = most useful) and they would recommend the course to another physicians. 19 scored the way of learning the management of cardiac critical situation as 5 of 5. 19 found the model as an adequate tool of training. 12 perceived an improvement of the ability to manage a pediatric cardiac critical patient.

**Conclusions:** Advanced medical simulation is a good tool to train pediatricians in cardiac critical care situations in a safe environment. Participants found the practice useful and they would recommend to another physicians. Evidence is growing to support medical simulation as the training tool of the future in pediatric cardiology. Further work is needed to prove the benefits in real situations.

#### P-290

##### **Surgical Correction of HOCM in Young Patients with Severe Hypertrophy**

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**Background:** The classic Morrow technique for HOCM in patients with extreme left ventricular hypertrophy and right ventricular obstruction is not effective. A new technique of HOCM surgical correction in patients with severe hypertrophy and septal myocardial fibrosis was proposed.

**Methods:** Conceptually, this approach offers a number of advantages: it affords the excision of the asymmetrically hypertrophied area of the ventricular septum without penetration into the left ventricle cavity; it avoids mechanical damage to the heart conduction system and aortic valve; and for surgeon, it improves visual inspection of the area to be resected. This excision was carried out on the right side of the IVS and not through the whole IVS thickness. 14 young patients with biventricular obstruction and severe hypertrophy (NYHA class 3) underwent this procedure. Episodes of ventricular tachycardia (VT) were registered in 8 patients. Ages ranged from 16 to 27 years. The follow-up period was  $34 \pm 7$  months.

**Results:** 12 patients were free of symptoms (NYHA class 1) and two patients had only mild limitations. The mean echocardiographic LVOT gradient decreased from  $86,9 \pm 12,7$  to  $10,1 \pm 2,3$  mmHg, the mean value of gradient in RVOT was reduced from  $41,7 \pm 5,1$  to  $4,3 \pm 1,3$  mmHg. Echocardiographically determined septal thickness was reduced from  $33,7 \pm 3,1$  to  $15,5 \pm 2,4$  mm. Sinus rhythm without block of His bundle right branch was noted in all patients after surgery. VT was not registered. None of the patients needed implantation of cardioverter-defibrillator.

**Conclusions:** This novel technique of HOCM surgical correction provides effective elimination of simultaneous LVOT and RVOT obstruction in patients with severe hypertrophy. A major advantage is that injures, in particular to the conduction system, are easily avoided.



## P-291

**Cerebral Magnetic Resonance Imaging before and after Neonatal Cardiac Surgery for severe Congenital Heart Disease**

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**Objectives:** To determine the influence of neonatal cardiac surgery on brain metabolism in neonates with congenital heart disease (CHD) using cerebral magnetic resonance imaging (MRI) and spectroscopy (MRS).

**Methods:** Cerebral MRI. (3T scanner), including single voxel spectroscopy in white matter (WM) and basal ganglia, was performed before and after neonatal cardiac surgery in fourteen patients with severe cyanotic CHD. Twelve patients were treated for transposition of great arteries (TGA) by arterial switch, 2 for hypoplastic left heart syndrome by Norwood or hybrid transcatheter-surgical palliation.

**Results:** Preoperative MRI was performed at median age of 6 days (range 1–12 d) before resp. 26 days (19–31 d) after neonatal cardiac surgery. Rashkind procedure was performed in 9 patients (75%) with TGA immediately after birth. Before surgery, all patients (100%) showed signs of generalized hypoxia with hyperintensity of the white matter (WM) on T2, with punctuate WM lesions in three patients (21%). Six patients (43%) showed hemorrhages either subdurally (n = 3) or in choroid plexus (n = 3). MRS was pathological in all patients with elevated brain lactate and decreased N-acetyl-aspartate (NAA) values. Only one patient (7%) with TGA had two small cerebral strokes after emergency shunt palliation due to closure of patent arterial duct. After surgery, hyperintensity of the WM decreased in most patients (54%). One patient (7%) had a new punctuate WM lesion. New hemorrhages occurred in 3 patients (21%) either subdurally (n = 1) or in choroid plexus (n = 2). Postoperative MRS could be analyzed in seven patients. The ratios of choline/creatine (pre 0.58 vs. post 0.46, p = 0.018) and myoinositol/creatine (1.74 vs. 1.48, p = 0.043) decreased from before to after surgery, whereas no significant difference was in ratio of lactate/creatine (0.28 vs. 0.22, p = 0.398). Neurological assessment showed muscular hypotonia (70%), but no focal neurological deficit or seizures.

**Conclusions:** Signs of generalized hypoxia in the WM are observed in all infants before cardiac surgery for CHD, but the extend of hypoxic brain alterations decreases after surgery. Hemorrhages are frequently observed, while cerebral strokes are rare in our population. None of the observed pathologies was accompanied by severe neurological symptoms. Further clinical follow up after one year is necessary.

## P-292

**Differences in Plasma B-type natriuretic peptide levels in children with univentricular heart malformation undergoing Fontan procedure**

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**Objective:** To determine plasma B-type natriuretic peptide (BNP) levels in children with complex types of univentricular heart malformation undergoing Fontan procedure.

**Methods:** In 70 patients with univentricular heart malformation BNP was determined before bidirectional Glenn anastomosis at an age (mean  $\pm$  SD) of  $0.4 \pm 0.2$  years before Fontan procedure at an age of  $2.8 \pm 1.5$  years (range 0.4–7.3) and after Fontan procedure at an age of  $9.7 \pm 4.6$  years (2.5–17.9).

**Results:** Children with univentricular CHD undergoing surgical staged palliation showed a significant decrease of plasma BNP levels undergoing Fontan procedure (mean BNP 116.4 pg/ml before Glenn anastomosis vs. 72.5 pg/ml before Fontan, (p < 0.05) and 43.3 pg/ml after Fontan). Comparing patients before Glenn anastomosis treated with modified Blalock–Taussig (BT) – Shunt showed significant lower plasma BNP level than patients treated with pulmonary artery banding (PAB) (BNP 86 pg/ml in patients treated with BT shunt vs. 176 pg/ml treated with PAB, p < 0.05) Comparing the morphologic type of systemic ventricle right ventricle morphology shows significant higher BNP plasma level than left ventricle morphology (BNP in left ventricle 59.1 pg/ml vs. 86.7 pg/ml in right ventricle, p < 0.05).

**Conclusions:** Plasma BNP levels decrease in patients with univentricular malformation undergoing Glenn anastomosis and Fontan due to volume relief. Patients with pressure overload of the systemic ventricle due to PAB are more affected compared to patients with volume overload due to BT-Shunt.

The effect of surgical treatment on plasma BNP levels highlights the usefulness of plasma BNP levels as diagnostic tool for follow up.

## P-293

**Hemodynamic consequences of postoperative inflammation after pulmonary valve replacement**

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**Objective:** Outcome of pulmonary valve replacement is typically excellent, but freedom from reoperation is variable and not predictable for the individual patient. There are some patients who develop early restenosis of the conduit valve or relevant conduit valve insufficiency. We speculated that the severity of inflammation immediately after the valve implantation may contribute to the early conduit failure.

**Methods:** From 8/2008 to 12/2010 pulmonary valve replacement by xenograft was performed in 42 patients (30 males, mean age 13.8 + standard deviation 6.6 years). Fifteen patients underwent a Ross procedure, 11 patients suffered from severe pulmonary insufficiency after Fallot-repair, and 14 patients required conduit replacement after previous conduit implantation for right ventricular outflow tract reconstruction. Hancock II<sup>®</sup>, Matrix-P/-P plus<sup>®</sup>, and Contegra<sup>®</sup> pulmonary valve conduits were used in 29, 11, and 2 patients, respectively. C-reactive protein (CRP) and white blood cell count (WBC) after surgery was retrospectively compared to the results of Doppler echocardiography during the follow-up after 3, 6, 12, and 24 months.

**Results:** CRP increased in all patients significantly within 48 hours after surgery to mean 151 + 69 mg/l. There was no significant difference in peak CRP value between the groups, neither between different surgical procedures nor between the different valves. WBC was mean  $12.6 + 3.7 \times 10^3 / \mu\text{l}$  and  $13.1 + 4.2 \times 10^3 / \mu\text{l}$  at the first 2 days. Mean follow-up was 12.4 + 8.7 months. Flow velocity in the pulmonary conduit at 3, 6, and 12 months after surgery was mean 2.0 + 0.5, 2.3 + 0.5, and 2.4 + 0.6 m/s, respectively. No or trivial conduit valve insufficiency was detectable in all patients during follow-up. Peak

CRP but not WBC was significantly correlated with Doppler peak flow velocity in the pulmonary conduit at 3 months after surgery ( $r = 0.33$ ,  $p = 0.03$ ).

**Conclusion:** Postoperative CRP level is correlated to the flow velocity in the conduit valve 3 months after replacement. However, long-term follow-up is necessary to answer the arising question whether postoperative inflammation is also negatively correlated to freedom from reoperation.

#### P-294

##### **Need of ECMO in a reversible life-threatening plastic bronchitis 8 years after Fontan completion**

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**Introduction:** Plastic bronchitis is a known but rare complication in patients with failing Fontan circulation, because of high central venous pressure, elevated transpulmonary pressure or pump failure. Bronchial casts with rubber-like consistency develop in the tracheobronchial tree and cause airway obstruction. The patients develop wheezing and desaturation and in case of ventilator therapy they need high mean airway pressures for oxygenation and therapeutic bronchoscopy to remove the casts.

**Case:** We report of a 12 year old boy in good clinical condition 8 years after total cavopulmonary connection. In a routine echocardiography an enlargement of the ascending aorta and aortic valve regurgitation was noticed without clinical symptoms. After replacement of the aorta ascendens and aortic valvuloplasty he showed a hard postoperative course with life-threatening lung haemorrhage, what was treated by transfusion of fresh frozen plasma and recombinant factor seven. Some days later he developed severe plastic bronchitis of the left lung. The patient got too hypoxic and hemodynamically unstable under high mean airway pressure for therapeutic bronchoscopy, so we took him on extracorporeal membrane oxygenation (ECMO) and removed the bronchial casts in stable condition. After diagnosing high degree of aortic valve regurgitation in the cath lab, the aortic valve was replaced. Two days later after normal bronchoscopy we weaned the patient from ECMO successfully. One year after the operation he is still free of plastic bronchitis and again in good clinical condition.

**Conclusion:** Almost one third of the reported Fontan patients with plastic bronchitis died of respiratory failure within one year, the rest suffer on recurrent episodes of respiratory compromise under different therapy (e.g. sildenafil, inhalation of recombinant tissue plasminogen activator). In case of an acute postoperative problem in Fontan circulation the occurrence of plastic bronchitis seems to be reversible, so each therapeutic approach, ECMO included, is warranted.

#### P-295

##### **Ultrasound Guided Insertion of Bi-Caval Dual Lumen Catheter for Venovenous Extracorporeal Membrane Oxygenation**

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**Introduction:** The new Avalon Elite<sup>®</sup> Bi-Caval Dual Lumen Catheter is the first single kit veno-venous ECMO cannula licensed for infants, children and adults.

It is designed to increase the efficiency of blood gas exchange by simultaneously removing desaturated blood from both the superior vena cava (SVC) and the inferior vena cava (IVC) and returning reoxygenated blood to the right atrium.

Proper positioning of the Bi-Caval Dual lumen ECMO cannula is imperative for optimal circuit function. Imaging is therefore mandatory during cannulation.

**Methods:** The Bi-Caval Dual Lumen ECMO cannula is placed using the Seldinger technique from the right internal jugular vein. It is the first ECMO cannula which must be placed into the IVC and have the reinfusion port positioned in the right atrium. Image guided placement is therefore essential.

**Results:** The Bi-Caval Dual Lumen single kit veno-venous ECMO cannula has been used in our institution in 71 Neonates, 13 Children and 183 Adults.

**Conclusion:** Echocardiography has been most effectively used and is our method of first choice for dual lumen ECMO cannula placement in newborns and in the emergency setting when there is not time to move the patient to the operating room for fluoroscopy. Fluoroscopy is the best imaging modality in larger children and adults.

#### P-296

##### **Aortopulmonary collaterals in neonates with d-TGA – clinical significance after arterial switch operation**

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**Objective:** In patients with d-TGA enlarged bronchial arteries or major aortopulmonary collateral arteries (MAPCA) are common. Although clinically silent, the cardiac volume overload after surgical repair can lead to longer mechanical ventilation, inotropic use and longer hospital stay. In addition, copious flow returning from these vessels to the left atrium can impair the view on the surgical field during cardiopulmonary bypass surgery.

**Methods:** We performed a 4-year retrospective monocentric study analyzing early clinical data in patients after arterial switch operation with MAPCAs complicating the postoperative course.

**Results:** In our institution 44 arterial switch operations were performed from 01.01.2007 to 31.12.2010. Postoperatively 17 (39%) patients needed a cardiac catheterisation due to known anomaly of the coronary arteries, pathologic signs for ischemia in ECG, or prolonged postoperative course. 10 of these 17 patients (59%) had one or more MAPCAs, and 8 required transcatheter coil embolization. There was no significant difference in gender and age. Catheterisation in patients with a hemodynamically relevant MAPCA was done between 7–53 (mean 18) days after switch operation without any major complications. Postoperative mean intubation time was 9.5 days, catecholamine use 9 days, and ICU stay 15 days. Overall hospital stay in patients with MAPCA coiling after arterial switch operation was 26 (mean, range 15–81) days. One patient with a hydrocephalus internus requiring a ventriculo-peritoneal shunt after cerebral hemorrhagia stayed in hospital for 81 days. In all 8 patients cardiac catheterisation resulted in complete MAPCA closure, but one patient required an additional operation for stenosis of the left coronary artery.

**Conclusions:** Hemodynamic relevant MAPCAs are quite common in patients after surgical repair of d-TGA. They can be large enough to cause pulmonary volume overload with the consequence of prolonged and complicated postoperative course. Therefore, when

facing this situation, an early postoperative work-up in the cathlab should be considered.

#### P-297

##### **Right ventricular hemodynamics are favored when the Rastelli or arterial switch procedure in double discordance is combined to a hemi-Mustard and bidirectional Glenn**

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**Introduction:** Anatomic repair of double discordance has become a useful surgical strategy with potential advantages over conventional surgical repair. In the presence of dextrocardia and situs solitus a complex Senning or Mustard operation is technically more demanding. In such patients we prefer to baffle the vena cava inferior to the right ventricle (hemi-Mustard) combined with a bidirectional Glenn. Other indications to perform a hemi-Mustard/Glenn instead of a complete atrial switch are RV hypoplasia and when a Rastelli procedure is performed. (where the LV-Ao tunnel and the RV-PA conduit negatively alter RV volume and function)

**Methods:** Between 2004 till 2009 hemi-Mustard/Glenn with Rastelli (n = 6) or arterial switch (n = 1) procedure were performed in 7 patients. Median age was 2.9 years (range 1.4–9.1 years). Atrial situs solitus was present in 6 patients and situs inversus in one. All but one with situs solitus had dextro- or mesocardia and the one with situs inversus had levocardia. Previous procedures were PA banding (n = 1), Glenn shunt (n = 1), systemic to pulmonary shunt (n = 4). Two patients had 2 systemic to pulmonary arterial shunts.

**Results:** One patient died postoperatively of Aspergillus sepsis. There was no late mortality. Two patients received a permanent pacemaker, one of them required ablation for atrial flutter. This patient had a RV to PA reoperation 6 years later. The latter is in New York Heart Association (NYHA) class II, the other 5 patients are in NYHA class I. At midterm follow up of mean 4.5 years there are no residual defects.

**Conclusions:** Hemi Mustard/Glenn is technically easier than full atrial switch when the ventricular mass is on top of the atria (situs solitus with dextrocardia or situs inversus with levocardia). Furthermore we strongly believe that addition of a Glenn shunt favors right ventricular hemodynamics especially when a Rastelli procedure is performed. Midterm follow up results are good.

#### P-298

##### **Value of Postoperative Hyperglycemia in Prediction of Low Cardiac Output State after Open Cardiac Surgery in Children**

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**Background:** Hyperglycemia is common in critically ill children including those who had repair of different types of congenital heart defects (CHD). Value of hyperglycemia in prediction of outcome in these patients is controversial.

**Methods:** During February 2009–January 2010, 78 consecutive children underwent repair of CHD using cardiopulmonary bypass. Blood sugar levels were checked every 6 hours for the first 48 hours after ICU entry and values above 150 mg/dl

considered hyperglycemia. Low cardiac output state (LCOS) defined as need for any inotropes more than 10 mcg/kg/min of dopamine for maintaining hemodynamic stability. Then correlation of postoperative hyperglycemia with LCOS and death in these patients studied.

**Results:** Median age was 15.5 months (7 days–17 years) and median weight 8.8 kg (3–66 kg). 24 patients (30.8%) developed LCOS and 4 (5.1%) died. Mean BS in 1st and 2nd postoperative days were 161.98 and 119.95 mg% respectively. In the first 24 hours, 44 patients (56.4%) and in the second day 6 (7.7%) developed hyperglycemia. There was no significant correlation between postoperative hyperglycemia and sex, age, weight, type of CHD, RACHS complexity score and duration of mechanical ventilation. Hyperglycemia in day 1 was correlated with number of inotropes ( $p = 0.005$ ) and in day 2 with bypass time ( $p = 0.004$ ). In the first postoperative day, 40.9% of the patients with hyperglycemia developed LCOS compared with 17.6% in those without hyperglycemia ( $p = 0.027$ ) [OR: 3.23, 95% CI: 1.1–9.3]. Hyperglycemia in second day was not accompanied with risk of LCOS ( $p = 0.069$ ), but it had a significant correlation with risk of death, 33.3% vs. 2.8% in those without hyperglycemia ( $p = 0.020$ ) [OR: 17.5, 95% CI: 1.9–158.5]. Mixing types of CHD ( $p = 0.007$ ) and pump time ( $p = 0.001$ ) were independent risk factors for LCOS and death. ROC curve analysis showed that BS > 134.8 mg/dl in the first postoperative day and bypass time > 68.5 min were cut off points for development of LCOS and death.

**Conclusion:** In this study hyperglycemia in the first postoperative day was common and a predictor of LCOS and in the second day, although not common, it was a predictor of death after open cardiac surgery in children.

#### P-299

##### **Early Predictors of Neurological Outcomes after Pediatric Heart Surgery**

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**Introduction:** Brain injury after pediatric heart surgery requiring cardiopulmonary bypass (CPB) and deep hypothermic circulatory arrest (DHCA) is common. Early identification of neurological damage is difficult and a clinically-available system to detect such injuries would be a significant advance. Near-infrared spectroscopy (NIRS) has been used to measure brain regional oxygenation saturations (RSO<sub>2</sub>) and various serological markers of brain injury (neuromarkers) have been proposed. However, a complete analysis of the ability of these systems to detect injuries that affect neurological outcome after heart surgery has not been performed.

**Hypothesis:** RSO<sub>2</sub> and neuromarkers are associated with adverse neurological outcome.

**Methods:** Prospective study of children (n = 48) undergoing heart surgery with CPB and/or DHCA. Brain RSO<sub>2</sub> was measured at baseline, continuously during surgery and the first 16h postoperatively. Neuromarkers (neuron-specific enolase (NSE), S100β and brain-derived neurotrophic factor (BDNF)) were measured at baseline, immediately after surgery and at 16h postoperatively. Neurological outcomes were defined as new abnormality on (i) examination, (ii) imaging or (iii) EEG within the first 3 months after surgery. Data presented as mean ± SEM unless otherwise specified.

**Results:** Median age was 8 m [5 d–17 y], 15 were neonates and 19 had abnormal neurological outcome (39.6%; 6 seizures,



7 abnormal 3 m exam). Percentage(%) of time with  $RSO_2 < 40\%$  during surgery or postoperatively ( $12.3 \pm 2.4$  v.  $5.4 \pm 1.9$ ,  $p = 0.03$ ;  $25.5 \pm 8.9$  v.  $6.9 \pm 3.5$ ,  $p = 0.02$ , respectively) and BDNF (in pg/ml) immediately after surgery ( $4460 \pm 1192$  vs.  $2539 \pm 485$ ,  $p = 0.001$ ) were significantly increased in children with abnormal neurological outcomes. In multivariate analysis, percentage of time with  $RSO_2 < 40\%$  during surgery and postoperatively and BDNF were independently associated with outcome.

**Conclusion:** Bedside neurological monitoring of  $RSO_2$  and novel neuromarkers may be helpful in assessing neurological injuries after pediatric heart surgery. A comprehensive study to determine the role of these modalities in neurological monitoring is needed.

### P-300

#### First Experience of Nikaidoh Operation

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**Objectives:** The choice of surgical management for patients with transposition of the great arteries, ventricular septal defect, and pulmonary stenosis is very difficult. The Rastelli operation and REV procedure are traditional surgical procedures, but their long-term results are not optimal. We would like to present our first experience using aortic translocation and biventricular outflow tract reconstruction for these patients and early post-operative period.

**Methods:** During 2010 year 6 patients have undergone aortic translocation and biventricular outflow tract reconstruction for the management of transposition of the great arteries, ventricular septal defect, and pulmonary stenosis at our institution. All patients had ventriculoarterial discordance, 4 of them had double-outlet right ventricle. An inlet ventricular septal defect was present in 3 patients.

The median age at operation was 20 months. Three patients had previous palliative procedures. The surgical technique used was the Nikaidoh procedure.

**Results:** There was no hospital death. The median intensive care unit stay was 18,5 days. At a median follow-up of 7 months, all patients are alive. One reoperation was performed in patient because of coronary artery problems.

**Conclusions:** Nikaidoh operation is a valuable surgical option for the surgical management of patients with transposition of the great arteries, ventricular septal defect, and pulmonary stenosis.

### P-301

#### Dilatable Banding of the Pulmonary Artery as An "open end" Palliation in patients with a Right systemic Ventricle

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**Introduction:** A morphologically right systemic ventricle (rSV) (post atrial switch, congenital corrected transposition (ccTGA)) is prone to develop failure and progressive tricuspid regurgitation (TR). Subgroups with (sub)pulmonary stenosis do better. The aim of this study was to assess the value of (dilatable) banding as an "open end palliation" in these patients.

**Patients and methods:** Single centre retrospective study. Twenty patients were banded: 5 late after atrial switch, age 8 years (5–12y), all fixed band. Fifteen patients with ccTGA: 6/15 had large VSD, age 2 months (1–7m); in 9/15 for prognostic reasons

at 4.5 years (8 months–15 years; dilatable band in 6). The dilatable band was established using a 2 mm synthetic cord, no knot, overlapping ends fixed by multiple vascular clips; the band can be adjusted percutaneously for somatic growth. The evolution of TR, biventricular function, gradient of the banding and need for interventions was assessed.

**Results:** Median follow-up 6 years. The median gradient evolved from 40 mmHg early PO to 55 mmHg when last seen. In patients with TGA after Senning, TR and the rSV function did not further deteriorate.

In patients with ccTGA a significant improvement of TR and rSV function is observed. When PA banding was done for prognostic reasons, no deterioration of the TV or rSV function is seen in these patients. Till present in only one patient a balloon dilatation of the banding needed to be done due to progressive cyanosis. None of the patients in both groups needed further surgical interventions (no need for double switch, no valvular repair, no pacemaker, no transplantation) till present.

**Conclusion:** Banding of the pulmonary artery in patients with rSV is a safe procedure with a good effect on the RV function and tricuspid regurgitation. Long term follow-up is needed but these preliminary data compare favourable to the natural history or other strategies.

Our current strategy in selected patients with rSV is to perform a dilatatable PA banding for prognostic reasons preferably before puberty as an "open end palliation".

### P-302

#### Pre-extubational BNP helps calculating post-extubational risks in patients with heart failure. A study in newborns after switch operation

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**Introduction:** Extubation of patients with heart failure may put them at risk for aggravation. Heart failure is associated with increased ventricular afterload. As brain natriuretic peptide (BNP) rises in the presence of increased afterload, it is supposed to rise after extubation. The aim of this study was to evaluate pre-extubational BNP as a predictor for post-extubational circulatory complications due to elevated afterload.

**Methods:** In 60 newborns after switch operation with left heart insufficiency, circulatory, respiratory, metabolic parameters and BNP were measured before and after extubation under conditions of minimal handling and unchanged treatment. They were compared for different pre-extubational BNP levels and BNP post-extubational dynamics. A BNP cut-off point was calculated for the prediction of post-extubational complications.

**Results:** BNP increased from pre-extubational 554.5 to post-extubational 1165 pg/ml. Clinical data remaining unchanged, pH and base excess as substrata for reduced cardiac output due to higher afterload decreased from 7.41 to 7.38 and from 1.5 to  $-0.05$  mval/l, respectively ( $p < 0.001$ ). Sensitivity for pre-extubational BNP as predictor of base excess decrease was 80.4%, its specificity 92.9%, with a cut-off point at 384 pg/ml. Absolute BNP increase was higher if pre-extubational BNP was higher ( $r = 0.77$ ), relative increase was highest for pre-extubational BNP under the cut-off point. Base excess decreased significantly for BNP beyond the cut-off but not below.

**Conclusions:** BNP measured before extubation can help to screen patients at risk for important changes in left ventricular afterload and subsequent signs of heart insufficiency after extubation.



## P-303

**Isolated Supravalvular Aortic Stenosis – Is a Coexistent Pulmonary Stenosis a Predictor of Unfavourable Outcome?**

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**Objective:** Certain factors have been implicated as indicators for increased risk of reoperations in children with supravalvular aortic stenosis (SVAS) including younger age at first operation, diffuse form of disease, coexistent valvular aortic stenosis and pulmonary stenosis (PS). According to the literature, a relatively high percentage of SVAS patients need a surgical or interventional relief of PS. Bilateral outflow tract obstruction is also known to increase the mortality risk in Williams Beuren Syndrome (WBS) patients. We sought to evaluate whether the presence of PS influences the rate of reoperations and mortality in patients with SVAS.

**Methods:** We identified the patients with isolated SVAS from our surgical database. The patients with a multi-level aortic stenosis as well as the patients with concomitant procedures were excluded from this study. Follow-up was conducted between 2008 and 2010 and is 100% complete.

**Results:** Twenty-six patients underwent surgical correction of SVAS between 1974 and 2006. The patients were operated at the age between 6 months and 26 years (median 8.8 years). Seventeen patients (65%) were diagnosed with WBS. Six patients (17%) had a diffuse form of SVAS and 10 (39%) had a diagnosed PS. No patient had a surgical or interventional procedure for PS during follow-up. There was no statistically significant correlation between PS and WBS ( $p = 0.302$ ) or diffuse form of SVAS ( $p = 0.128$ ). Patients with PS were operated at younger age ( $4.4 \pm 2.7$  compared to  $12.2 \pm 7.1$  years;  $p = 0.028$ ). Median follow-up time was 14.6 years. Overall mortality was 11.5%. One patient with preoperatively severely reduced LV-function died 27 days postoperatively. Two late deaths occurred 7 and 10 years after the initial operation. Reoperations were required in 4 patients (15%), 4–19 years after the original operation, due to stenosis of the aortic arch, supravalvular restenosis or poststenotic aortic dilatation. PS was found to be a risk factor for reoperation ( $p = 0.005$ ) as well as for the combined end point reoperation/death ( $p = 0.003$ ).

**Conclusions:** PS in patients with SVAS is a risk factor for reoperations in the aortic region and might be considered an indicator of the severity of the arterial disease and a predictor of an unfavourable outcome.

## P-304

**Perioperative cytokine release and its effects on the early postoperative course in patients undergoing extracardiac Fontan operation (ECFO) in an off-pump technique compared to CPB-supported ECFO**

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**Objective:** Cardiopulmonary bypass in pediatric heart surgery is still assumed to be a major cause of systemic inflammatory response syndrome (SIRS).

We investigated perioperative cytokine release and its effects on the early postoperative course in patients undergoing extracardiac Fontan operation (ECFO) in an off-pump technique compared to CPB-supported ECFO.

**Methods:** In the last consecutive 22 of the total of 138 patients (off-pump:  $n = 11$ , on-pump CPB:  $n = 11$ ) undergoing ECFO in our institution, plasma concentrations of IL-1alpha, IL-1beta, IL-6, IL-8, IL-10, TNF-alpha and MCP-1 were measured pre- and perioperatively, immediately after CPB and at 4h and 24h postoperatively. Median age of patients studied was 4.0 (1.7–12.3) years and median weight 12 (9.8–53.8) kg. Clinical signs of cardiac, pulmonary and renal dysfunction were evaluated.

**Results:** There were no differences between the two groups regarding preoperative hemodynamics and anthropometric data or cytokine levels except for IL-1alpha ( $p = 0.028$ ). Plasma levels of all mediators were below reference values preoperatively and rose during surgical procedure in both groups. There were significantly higher values of IL-1alpha ( $p = 0.016$ ), IL-8 ( $p = 0.019$ ), IL-10 ( $p = 0.023$ ) and TNF-alpha ( $p = 0.016$ ) in the CPB group during surgical procedure. All parameters peaked at the 4hour postoperative measure with IL-6 reaching 241 times, IL-8 34 times and TNF-alpha 1.7 times the preoperative levels. In the CPB group, plasma levels were significantly higher for IL-1a ( $p < 0.001$ ) and IL-10 ( $p = 0.04$ ) 4h postoperatively. Twenty-four hours after operation no differences between groups were found. However, all parameters were still above preoperative levels.

High perioperative cytokine plasma levels (IL-1alpha, IL6, IL8, IL10 and MCP1) correlate significantly with higher postoperative creatinine level as well as pleural effusion volume and positive fluid balance 24 and 48h postoperatively.

High levels of IL-8 correlated significantly with prolonged mechanical ventilation ( $>24$ h), ICU stay and duration of pleural effusions.

**Conclusions:** The results emphasize the contributing effect of CPB in cytokinemia and systemic inflammation that correlates with increased early postoperative requirement of fluid resuscitation and inotropic agents. Completion of Fontan circulation using off-pump technique for extracardiac conduit may improve early postoperative outcome.

## P-305

**Oral sildenafil early after Fontan operation improves postoperative outcome**

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**Objective:** Elevated pulmonary vascular resistance (PVR) is a risk factor for early Fontan failure. Oral sildenafil was been increasingly applied in recent years in addition to inhaled nitric oxide (NO) in an attempt to reduce PVR and improve outcome. To evaluate the efficacy of this medication we analyzed retrospectively our experience with NO and oral sildenafil after extracardiac Fontan operation (ECFO).

**Methods:** A total of 138 patients underwent ECFO at our institution (1997–2010). Preoperative data were in median: age 4.5 years, weight 16 kg, oxygen saturation 82%, Hb 16.0 g/dl. Pts. were selected for Fontan with median mean pulmonary artery pressure (mPAP) of 10 mmHg, median Nakata index of  $228 \text{ mm}^2/\text{m}^2$  and median lower lobe index of  $143 \text{ mm}^2/\text{m}^2$ . NO was started early postoperatively if the mPAP was 16 mmHg or more. Two main groups (requiring inhaled NO ( $n = 50$ ) or not ( $n = 88$ )) were comparatively analyzed and subgroups with oral sildenafil were added. Sildenafil was given from the 1st postoperative day and continued after discharge in the last 15 consecutive pts., who required NO, and in 7 with preoperatively elevated mPAP (14–17 mmHg) without necessity for inhaled NO.

**Results:** There were no differences in the preoperative data between the pts. who required inhaled NO and those who did not. The total mortality was 9 pts. (6.5%) and correlates with NO requirement ( $p < 0.001$ ). Longer CPB duration ( $>1$  h,  $n = 94$ ) correlates with necessity of NO therapy ( $p = 0.006$ ). Patients requiring NO had worst postoperative course with elevated mPAP (median 14 vs. 12 mmHg,  $p < 0.001$ ), prolonged ventilation (median 10 vs. 86 h,  $p < 0.001$ ) and ICU stay (median 2 vs. 7 days,  $p < 0.001$ ) and higher incidence of prolonged effusions ( $>10$  days,  $p = 0.004$ ). Patients who received sildenafil in addition to NO had a better postoperative course with lower incidence of prolonged effusions compared with isolated NO group ( $p = 0.002$ ).

**Conclusion:** Necessity of inhaled NO remains a significant factor, indicating early Fontan failure. Prolonged CPB seems to increase the PVR with NO requirement. Additional use of oral sildenafil improves postoperative outcome and reduces the incidence of pleural effusions. Further prospective and randomized studies are necessary.

### P-306

#### Levosimendan in dilated cardiomyopathy and refractory cardiogenic shock in children

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**Introduction:** Levosimendan is a new calcium sensitizer and K-ATP channel opener. The documentation regarding this drug is one of the largest ever on the safety and efficacy of a new pharmacological agent in acute heart failure syndromes in adult population [1]. Its use in paediatric is limited to successful weaning from biventricular mechanical support in case reports or small trials conducted in the immediate postoperative period [2]. We report our experience of using Levosimendan during refractory cardiogenic shock (RCS) in infants.

**Patients and methods:** Four infants aged 2–24 months and suffering from hypokinetic dilated cardiomyopathy were included in this study. All presented with uncontrolled RCS (LVEF  $< 20\%$ ) despite conventional inotropes treatment. Lev was intended as a last resort before ECMO. The effectiveness of treatment was monitored by measuring the echocardiographic left ventricular ejection fraction (LVEF) and plasma BNP assay before and 8 days after administration. A total of 15 injections of Lev were realized and studied.

**Results:** Mean LVEF before and 8 days after administration of Levosimendan significantly increase from  $19.75\% \pm 1.7$  to  $33 \pm 2.95$  ( $p < 0.01$ ) and mean BNP level decrease from  $2267 \text{ pg/ml} \pm 518$  to  $1673 \pm 372$  ( $p < 0.08$ ). These children could benefit from additional treatments of Lev in order to wean amines and defer the circulatory support. In one of these, a total of 6 cycles were necessary without the use of circulatory support for an improvement. In the 4 patients studied, the outcome was favorable.

Figure 1: the individual changes in LVEF and BNP after each injection of Levosimendan

**Conclusion:** During the refractory cardiogenic shock of the child with hypokinetic dilated cardiomyopathy, levosimendan may improve myocardial function allowing weaning of conventional inotropes and circulatory support. Re-injection may also be necessary. Randomized studies with larger number of patients are needed to confirm these very encouraging results.

1. Kota B, et al. J Cardiovasc Pharmacol Ther. 2008; 13: 269–78.
2. Eriksson HI, et al. Ann Thorac Surg. 2009; 87: 448–54

### P-307

#### Chylothorax – do standard management techniques affect prolonged pleural drainage in post-operative cardiac surgery patients?

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**Introduction (or Basis or Objectives):** Chylothorax is an uncommon complication of paediatric heart surgery (approx. 1–2%). The diagnostic criteria for this condition are the presence of chylomicrons, triglyceride concentration, total cell count, and lymphocyte percentage. The aims of this project were to investigate the correlation between such investigation results and duration of drainage and length of hospital stay, and to determine the impact of interventions on these outcomes.

**Methods:** Data was reviewed from a three-year period (2007–10) from patients aged  $< 16$  who had been tested for suspected chyle in pleural effusion post-cardiac surgery, and had survived until discharge. Key outcomes were duration of drainage (DOD) and length of hospital stay (LOS). 76 patients were allocated to groups (positive, borderline and negative) based on the results of protocol based investigations.

**Results:**

	Positive	Borderline Group A ‡	Group B ‡
No. of patients	33	5	20
Age (median (range))	8 months (10 days – 10 years)	1 year (7 days – 8 years)	1 year (25 days – 7 years)
DOD (median)	10	12	9.5
LOS (median)	20	46	17
<b>Positive and borderline patients only</b>			
	Treated	Untreated	
DOD (median)	12	7	
LOS (median)	23	15	
	Negative	p-value	
No. of patients	18		
Age (median (range))	3.5 years (1 year – 10 years)		
DOD (median)	14	0.638	
LOS (median)	19.5	0.530	
DOD (median)	0.006		
LOS (median)	0.031		

‡ The borderline group – in which patients were negative for chylomicrons – was subdivided into Group A (patients who were positive for the combination of triglyceride concentration, total cell count, and lymphocyte percentage) and Group B (patients who were positive for only one or two of these).

There were no significant differences between groups in terms of DOD or LOS. Low fat diet and medium chain triglyceride substitution did not show a significant impact on these outcomes. TPN and octreotide were associated with poorer outcomes ( $p = 0.02$ ).

**Conclusions:** Differentiating chylous from non-chylous effusions with corresponding management strategies has little impact on either duration of drainage or length of stay. TPN and octreotide appear to be associated with longer DOD and LOS; however, it seems more likely that TPN and octreotide are utilised late in the process. Given that interventions themselves may lead to adverse effects and complications, and in some cases are based on anecdotal evidence, we feel a randomised trial of active vs. expectant management is indicated. On the basis of these findings in our department, we have adopted a laissez-faire approach to chylothorax by delaying

interventions for one week post-diagnosis (as we found that the median duration of drainage in untreated positive patients was 7 days). This approach has already proved successful in 2 cases.

### P-308

#### Sildenafil Treatment in the Postoperative Stage I and II in Univentricular Circulation

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**Background:** Sildenafil in the management of patients with univentricular circulation is less well defined and as yet has only been reported sporadically.

**Objectives:** Present our experience with sildenafil treatment in patients with Univentricular circulation, started in the immediate post-Norwood or Glenn Surgery.

**Methods:** We retrospectively studied 16 patients (mean age 5.03 months; 8 females; mean weight 4.78 kg), treated with oral sildenafil between January 2008 and December 2010 (Postop Stage I 4/16, Postop Stage II 12/16). Dosage (mg/kg/day), duration and side effects were recorded. Basal data in the immediate postoperative period and at 6 months after Glenn, or on stopping treatment was analysed if they achieved: 1) Respiratory improvement (mechanical ventilation withdrawal, supplementary oxygen, nitric oxid); 2) Clinical situation (improvement or maintenance of oxygen saturation); 3) Decrease in debit drains. We analyzed echocardiographic and hemodynamic parameters response in those who underwent catheterization. Pulmonary artery stenosis or other anatomic defects which could compromise pulmonary flow and posterior interventions to achieve hemodynamic or clinical status after treatment were also recorded.

**Results:** Mean initial dosage was 1.96 mg/kg/d (Range 0.5–4) and mean duration of treatment was 12.4 months. Escalating to the maximum dosage occurring within 48 hours in 13/16 (81,3%), mean maximum dosage 2.53. Side effects were observed in 2 patients, hypotension (100%), not requiring withdrawal in any case. The average number of days in which treatment was initiated after surgery was 6.17 days. At the time of initiation of treatment, 9/16 (56.2%) patients were on mechanical ventilation, 8/16 (50%) had nitric oxid, 4/16 had moderate to severe ventricular dysfunction. An improvement was observed on commencing in 10/16 patients, especially in respiratory evolution (62.5%). Patients who underwent intervention to optimize pulmonary flow post-treatment 11/16 (5/16 pulmonary artery stenosis, 4/16 collateral circulation) had a positive response in 9/11 (80%). Patients with worse ventricular function had a worse response ( $P = 0,046$ ). In patients who underwent catheterization pre and post treatment showed a tendency in decrease in mean pulmonary pressures (Median Pre-treatment 14 – Median Post-treatment 13).

**Conclusion:** Sildenafil can be used with favourable results in the post-operative setting of children who undergo Norwood and Glenn palliation.

### P-309

#### Survival and catheter interventions after Norwood surgery

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**Introduction:** Hypoplastic left heart syndrome and other functionally single ventricle malformations can be palliated with the Norwood procedure. The mortality and morbidity of the procedure is still considerable. We describe the results of Norwood surgery and the need for catheter interventions in patients after Norwood surgery in a single center.

**Methods:** From 1999 to 2009 all patients surgically palliated by the Norwood procedure were included in a retrospective study. In the early years the pulmonary blood flow was supplied by a modified BT-shunt or central shunt, which later changed to a right-ventricle to pulmonary-artery shunt. Obstructions in the pulmonary branches or aortic arch, even mild or moderate, were treated by catheter interventions.

**Results:** A total of 60 patients were palliated by the Norwood procedure, 19 received a BT/central shunt and 41 a RV-PA shunt. Early mortality, that is death within 30 days of surgery, was 13% ( $N = 8$ ; 4 BT/central, 4 RV-PA). Mean age at the Glenn procedure was 5.5 months (range 3–10) and at completion of the Fontan circulation 3.2 years (range 2.1–4.7). The 5-year survival after surgery was 69% (figure strait line) with a mean follow-up of 5.7 years (range 1.5–10.4). A total of 50 catheter interventions were performed in 24 patients surviving more than 30 days after surgery (46%), not including closure of fenestrations (24 patients). The 5 year intervention free survival after Norwood surgery was 36% (figure dashed line). In 13 patients (4 BT/central, 9 RV-PA) 16 aortic arch dilatations were performed, in one patient followed by placement of a stent. In 12 patients (2 BT/central, 10 RV-PA) diagnosed with a stenosis of the left or right pulmonary artery branch, 16 stents were implanted and in 6 of these patients 8 balloon dilatations were performed in a separate session. Eight collaterals were closed in 6 patients by coils or plugs and in one patient a balloon dilatation of vena anonyma stenosis was performed.

**Conclusion:** In this series of patients the long-term survival after Norwood surgery is good. During follow-up catheter interventions are often required to further optimize hemodynamics in this vulnerable group of patients.

